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EDITORIAL

RHEUMATOID ARTHRITIS

There has been great interest in rheumatoid arthritis and diseases thought to be related to it since the discovery a few years ago that cortisone and other steroids produce symptomatic improvement in these conditions. However, the earlier hopes about such therapy have not been fulfilled, and until the true cause of these disorders becomes established the chances of curative treatment becoming available are not very great.

At present the only reason for grouping the 'rheumatic' diseases together seems to be the common site of their primary lesions in the connective and supporting tissues. The onset of rheumatoid arthritis has been attributed to a number of predisposing or precipitating factors, but these theories have mostly been based on clinical impressions and on statistically uncontrolled studies. A controlled investigation a few years ago by the Empire Rheumatism Council [1] and other workers [2] indicated that a familial history of arthritis, exposure to cold, and peripheral circulatory disorders, occurred in a significant number of the cases.

There is no good evidence that focal sepsis is of any significance in the pathogenesis of rheumatoid arthritis, although the concept is by no means yet abandoned. There is considerable evidence against a streptococcal origin of the disease, despite the high titre of haemolytic streptococcal agglutinins present in the serum in active rheumatoid arthritis and the ease with which a chrome polyarthritis can be produced experimentally. It would appear that streptococci are of no significance in the pathogenesis of rheumatoid arthritis although streptococcal infection is a possible factor precipitating the onset of the disease or its recurrences. The streptococcal agglutinins in the serum are presumably non-specific and non-antigenic in nature. There is believed to be an abnormal globulin factor in the serum in rheumatoid arthritis which accounts for the tendency of such sera to precipitate spontaneously in saline, and which probably exaggerates the action of naturally-

VAN DIE REDAKSIE

MISVORMENDE GEWRIGSONTSTEKING

Sedert dit 'n paar jaar gelede ontdek is dat cortisone en ander steroïede die simptome van misvormende gewrigsontsteking en vermoedelik verwante siektes verlig, wek hierdie toestande groot belangstelling. Die vroeë verwagtings van hierdie terapie is egter nie verwesenlik nie en totdat die juiste oorsaak van hierdie kwale vasgestel word is die kanse vir helende behandeling maar skraal.

Die enigste rede waarom ‚rumatiek'-siektes tans saamgegroepeer word, is waarskynlik daaraan te danke dat in al hierdie kondisies die vernaamste letsels in die bind- en die stutweefsels voorkom. Misvormende gewrigsontstekingaanvalle word aan 'n aantal predisponerende of presipiterende faktore toegeskryf maar hierdie teorieë is grotendeels gebaseer op kliniese indrukke en studies wat nie statisties gekontroleer is nie. 'n Gekontroleerde ondersoek wat 'n paar jaar gelede deur die Empire Rheumatism Council [1] en ander werkers [2] ingestel is, het aan die lig gebring dat 'n familiegeskiedenis van gewrigsontsteking, blootstelling aan koue en ongesteldhede van die perifeerbloedsomloop in 'n aansienlike aantal gevalle voorkom.

Daar is geen grondige bewys dat haardbesmetting 'n veroorsakende faktor in die ontstaan van misvormende gewrigsontsteking is nie, alhoewel hierdie teorie nog glad nie op die lang baan geskuif is nie. Daar bestaan heelwat bewyse teen 'n streptokokke-oorsprong, ten spyte van die hoë titer van hemolitiese streptokokke-agglutiniene wat met aktiewe misvormende gewrigsontsteking in die serum gevind word en die gemak waarmee 'n uitgebreide chroomgewrigsontsteking eksperimenteel teweeggebring kan word. Oënskynlik speel streptokokke geen rol in die ontstaan van die siekte nie alhoewel streptokokkusinfeksie moontlik as presipiterende faktor in aanvalle of hernude aanvalle ageer. Die streptokokkus-agglutiniene in the serum is vermoedelik nie-spesifiek en nie-antigenies van aard nie. Dit word gemeen dat in misvormende gewrigsontsteking 'n abnormale globulienfaktor in die serum aanwesig is, wat verantwoordelik is vir die neiging van dié serum om in 'n soutoplossing spontaan neer te slaan, en wat waarskynlik die aksie verhoog van presipitiene vir streptokokkus- en pneumokokkus-fraksies wat natuurlik voorkom.

Gordon [3] het geredeneer dat 'n groep spesifieke virusse

occurring precipitins for streptococcal and pneumococcal fractions.

Gordon [3] has argued that a group of specific viruses with selective affinity for connective tissue may be the cause of the disorder in rheumatic diseases and, though it seems unlikely that the lesions of rheumatoid arthritis are due to a virus,[4, 5] this cause cannot as yet be definitely excluded.

The Empire Rheumatism Council in its investigation [1] found no significant relationship between psychic trauma and the onset of the disease. There are some who classify rheumatoid arthritis as a psychosomatic disorder, but most physicians do not regard the disease as having this origin although emotional disturbances are accepted as having a relationship to fluctuations in the disease. There is no evidence definitely to connect avitaminoses with the disease; a deficiency of ascorbic acid is more likely a result rather than a causal factor in rheumatoid arthritis. As regards endocrine dysfunction evidence is available that abnormal steroid metabolism may be present, but whether this is a result of the disease or a cause it is impossible to state at present. Although the adrenal steroids modify the reaction their effects are not specific and they have not yielded direct information on the aetiology of rheumatoid arthritis; their therapeutic effects have of course given a great stimulus to the study of the pathogenetic mechanisms.

The term 'collagen-vascular diseases' has been used for the group which includes rheumatoid arthritis, rheumatic fever, polyarteritis nodosa, and dermatomyositis. There is a non-specific disturbance involving connective tissues, especially the extracellular elements, which is observed microscopically as a form of 'fibrinoid' degeneration and a profound physicochemical alteration of the interfibrillar ground-substance and possibly of the scleroprotein fibres also. A knowledge of the nature of this basic change might throw light on the pathogenesis of these disorders. The subject is obviously one of great complexity. The evidence seems to support the view that the inflammatory and toxic features of rheumatoid arthritis result from primary disintegration of collagen. The concept of rheumatoid arthritis as one of the connective-tissue diseases does not however necessarily imply a unified aetiology for the whole group.

A lengthy and excellent review on the aetiology and pathogenesis of rheumatoid arthritis has recently been published,[6] in which not only the factors referred to above are discussed fully but also other aspects of rheumatoid arthritis, such as the question of hypersensitivity of the tissues and Selye's concept of disorders of adaptation. Our knowledge of rheumatoid arthritis has greatly advanced in recent years but the factors predisposing to or maintaining the disease have not been clearly established.

1. Empire Rheumatism Council (1950): Brit. Med. J., **1**, 799.
2. Short, C. A. *et al.* (1949): Ann. Rheum. Dis., **8**, 313.
3. Gordon, M. (1948): Lancet, **1**, 697, 740.
4. Levinsky, W. J. and Lansbury, J. (1951): Proc. Soc. Exp. Biol., **78**, 325.
5. Bauer, W., Clark, W. S. and Dienes, L. (1951): Practitioner, **166**, 5.
6. Dresner, E. (1955): Amer. J. Med., **18**, 74.

met 'n selektiewe affiniteit vir bindweefsels vir rumatiekkwale verantwoordelik mag wees en alhoewel dit onwaarskynlik is dat die letsels van misvormende gewrigsontsteking aan 'n virus te wyte is [4, 5] kan dit nog nie definitief as 'n oorsaak uitgeskakel word nie.

Die ondersoek van die Empire Rheumatism Council het geen noemenswaardige verwantskap tussen psigiese trauma en 'n aanval van hierdie siekte aan die lig gebring nie. Daar is diegene wat misvormende gewrigsontsteking as 'n psigosomatiese kwaal bestempel, maar die meeste geneeshere glo nie dat dit die oorsprong van die siekte is nie alhoewel dit aanvaar word dat daar 'n verhouding bestaan tussen emosionele steurings en wisselings in die siektetoestand. Dit is nie definitief bewys nie dat daar enige verband tussen hierdie siekte en 'n vitamiengebrek is nie; dit is meer waarskynlik dat 'n tekort aan vitamien C eerder 'n gevolg as 'n oorsakende faktor in misvormende gewrigsontsteking is. Wat die onreëlmatige werking van die buislose kliere betref is dit bewys dat abnormale steroïde-metabolisme aanwesig mag wees maar tot nog toe is dit onmoontlik om te konstateer of dit die gevolg of die oorsaak van die siekte is. Alhoewel byniersteroïde die reaksie beïnvloed is hul uitwerking nie spesifiek nie en verstrek hul nie direk inligting oor die etiologie van misvormende gewrigsontsteking nie; vanselfsprekend het hul terapeutiese gevolge die studie van die patogenetiese meganismes aangewakker.

Die benaming ‚kollageenvatsiektes' is vir dié groep gebruik wat misvormende gewrigsontsteking, rumatiekkoors, knopvormige slagaarontsteking en dermatomyositis behels. Daar is 'n nie-spesifieke steuring wat die bindweefsel, veral die buitesellulêre elemente, raak wat onder die mikroskoop as 'n vorm van fibrienagteruitgang en 'n groot fisies-chemiese verandering van die tussenfibrillêre grondstof (en moontlik ook van die skleroproteïenvesels) gesien word. Kennis van die aard van hierdie basiese verandering mag lig op die ontstaan van hierdie siekte werp. Hierdie onderwerp is ongetwyfeld 'n baie ingewikkelde een. Die gegewens blyk die opvatting te staaf dat die ontstekings- en vergiftigingseienskappe van misvormende gewrigsontsteking op die primêre disintegrasie van die kollageen volg. Die mening dat misvormende gewrigsontsteking 'n bindweefselsiekte is, behels nie noodwendig die opvatting dat die oorsaakleer vir die hele groep dieselfde is nie.

'n Breedvoerige en puik oorsig van die oorsaakleer en die siekte-ontstaan van misvormende gewrigsontsteking is onlangs gepubliseer,[6] waarin nie net die bogemelde faktore ten volle bespreek word nie maar ook ander aspekte van misvormende gewrigsontsteking soos byvoorbeeld die kwessie van oorgevoeligheid van die weefsels en Selye se opvatting van aanpassingsmoeilikhede. Ons kennis van misvormende gewrigstontsteking het in onlangse jare baie gevorder maar die faktore wat hierdie siekte in stand hou of daartoe predisponeer is nog nie duidelik vasgestel nie.

1. Empire Rheumatism Council (1950): Brit. Med. J., **1**, 799.
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POLIO VACCINE

The results of the great American experiment with the Salk polio vaccine were announced at the University of Michigan on 12 April, and from the messages which were published in the South African press the next day it appears that the hopes of the promotors of the mass test have been realized. As in all field tests of this kind, the interpretation of the results is a problem for statisticians. The controversy that arose out of the early results—and indeed the later results—of the use of B.C.G. as a prophylactic against tuberculosis will be remembered. The medical profession in South Africa are therefore compelled to wait until the official reports from America reach this country before they can come to final conclusions. These reports are expected to arrive shortly; copies may indeed be available here by the time this issue of the *Journal* is published.

The press messages report Dr. Thomas Francis, who is in charge of the evaluation, as saying that the vaccine was found to be 80 or 90 per cent effective in preventing paralytic polio, but other disconnected figures are given which cannot be interpreted without fuller details. It is stated that the results varied for the 3 types of poliovirus. The question of the duration of immunity has received attention and the messages include the statement that the effect was maintained with but moderate decline after 5 months. The vaccine was found to be very safe in use, the messages stating that only 0·4 of 1 per cent of the children injected suffered from minor reactions and that major reactions were almost entirely absent.

In view of these statements the official reports may be awaited with a high degree of confidence, for the mass experiment was designed and the results analysed in order to give trustworthy replies to the questions to which answers were wanted. They will be of particular interest in South Africa because the work of producing a polio vaccine similar to the Salk vaccine is far advanced at the laboratories of the Poliomyelitis Research Foundation where, although the vaccine has not yet been tested on human beings, enough of it is in stock to vaccinate the whole population of the Union. South Africa is one of the few countries in the world which have seriously attacked the task of producing a prophylactic polio vaccine.

The news about the polio vaccine naturally provokes comparison with anti-diphtheria inoculation. In both the routine is a series of 3 injections at intervals of a few weeks, with a possibility that 2 injections may be adequate, especially if later reinforced by a 'booster' dose. The practical problems therefore appear to be similar. The case for universal protective inoculation of children is not the same in the two diseases. In diphtheria the deaths that occur in the community when the disease is uncontrolled are far more numerous than ever happen from poliomyelitis. Poliomyelitis becomes epidemic at certain times and places, but at any one time it is in an endemic phase in most parts of the world, when the amount of mortality for years on end, and even the number of cases, may be practically negligible. The dread which polio inspires in the community when it becomes epidemic in a country is due to the disabling crippling that is the common sequela of a paralytic attack. The impression that this made on South Africa a few years ago was reflected in the public response to the appeal for funds to combat the disease.

It may be expected that in America, where the present mass experiment has been followed with enormous nation-wide interest, a relatively complete inoculation of the child population against polio will soon be under way, and the manner in which the prophylactic will be used in other countries will no doubt be influenced by the further experience which will thus be gained. The remarkable feature of immunization against the greater menace of diphtheria is that, in spite of its long-proven efficacy, in most parts of the world—South Africa included—public apathy has been sufficient to prevent the virtual abolition of diphtheria which a universal acceptance of inoculation in childhood would secure.

RADIOLOGICAL FEATURES OF NEUROFIBROMATOSIS

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Von Recklinghausen [1] in 1882 first established the true nature of the tumours in this disease. Lichtenstein and Jaffe [2] (1942) consider that the underlying cause is a fundamental genetic defect, but according to Willis [3] (1948) the absence of a family history is not unusual.

The primary characteristic of the disease is the development of cutaneous and subcutaneous tumours arising from the peripheral nerves; but there are, however, many other features, which are summarized by Friedman [4] (1944) as (1) pigmentation of the skin, (2) mental deficiency, (3) involvement of the central nervous system, (4) congenital developmental defects, and (5) alterations in the skeleton.

Brooks and Lehman [5] (1924) were apparently the first to draw attention to local bone changes, such as subperiosteal and cortical 'cysts'. A condition of 'neurofibromatous elephantiasis' was found in 2 of the 7 cases they described. According to Weber [6] (1930) this term

is applied to an extreme degree of plexiform neuroma and pachydermatocele of the soft parts of an extremity. He expresses the opinion that more or less diffuse neurofibromatous thickening of the periosteum is likely to be present in these cases to account for the bony hyperplasia. He thinks that the latter is intimately connected with excessive blood supply. The incidence of skeletal changes in neurofibromatosis is quoted as 7% by MacKenzie [7] (1950) and Friedman [4] (1944), though Holt and Wright [8] (1948) reviewing 127 cases over a period of 13 years (1934-1947) find the incidence higher. The bony changes are well summarized by MacKenzie. He points out that the erosive defects are frequently due to pressure, for they bear a close relationship to the soft tissue lesions. He states that the disorders of bone growth include both atrophy and hypertrophy, sometimes concurrently at different sites of the same bone

Moore [9] (1941) expresses the opinion that a growth distortion or faulty control of growth accounts for the occurrence of hypertrophy and under-development. This growth disturbance is also the underlying factor in the pseudarthrosis cases, usually seen in children.

According to Fairbank [10] (1951) the neurogenic origin of the subperiosteal bone lesions is readily acceptable, but this is not so with those that are endosteal. Nevertheless the typical fibrosis with 'whorls' of cells has been found in some lesions apparently endosteal. MacKenzie [7] holds that the intraosseous cystic areas are due to lesions of nerve endings which penetrate into the bone with the nutrient vessels, but does not submit any proof.

Friedman [4] reported one case where biopsy from a femur showed growth in marrow spaces which closely simulated neurofibromatous structure, suggesting that nervous tissue must be present in the bone marrow.

A kyphoscoliosis is a frequent finding in this disease. The deformity is usually most marked in the lower thoracic region. Miller [11] (1936) said it was present in 45% of cases. The cause of the scoliosis is controversial. Gould [12] (1918) thought it was due to bone softening. Brooks and Lehman [5] held that it was caused by neurofibromata of spinal nerves. Moore [9] associated it with growth distortion. MacKenzie [7] thought that it was due either to an associated congenital defect or to asymmetrical erosion of a vertebra.

According to Fairbank [10] the scoliosis usually shows a tendency towards progressive increase of the curves. Paraplegia may develop in cases with severe scoliosis. This may occur when a spinal root is the seat of a neurofibroma.

Spondylolisthesis was first reported in this condition in 1950 by McCarroll.[13] He described 4 cases, and postulated that it was due to congenital pseudarthrosis of the pedicle. He said that the causal relationship to neurofibromatosis was not proven.

McCarroll also quotes cases with marked vascular changes, notably haemangiomata of the diffuse flat type, a plexiform type with dilated veins and lymphatic oedema of hypertrophied extremities.

The occurrence of secondary malignant changes is mentioned by Moore,[9] and MacKenzie [7] states that sarcomatous change may occur in more than one fibroma.

CASE REPORTS

Case 1

A Bantu female aged 29 years, admitted on 14 April 1954 complaining of abdominal pain and swelling of 6 months' duration. Jaundiced for 2 weeks. Numerous lumps all over the body, which started from a single small lump on the left ear—8 years ago.

Examination. Numerous soft, subcutaenous, well-defined, non-tender masses, varying from 2 mm. to 10 cm. over entire body surface. Large swelling of similar nature on lateral side of right thigh. Patient wasted, with marked jaundice and pallor of mucous membranes. In the abdomen: extensive ascites, spleen 5 inches below rib margin, no other masses felt. Remaining systems normal.

Diagnosis: Hypersplenism, haemolytic anaemia and neurofibromatosis.

Special Investigations were made relevant to the hypersplenism and the haemolytic anaemia, but they have no bearing on the neurofibromatosis.

X-Ray. No abnormality discovered in the chest or spine. *Pelvis and Femora:* There were exostoses and some bone proliferation on the lateral aspect of the right ilium, which was flattened. There was a rounded ossification about the size of a hazel nut just above the upper border of the greater trochanter of the right femur. The greater trochanter was flattened on its lateral aspect and showed areas of increased density. It was thought probable that these bone changes were related to the soft-tissue mass on the lateral aspect of the upper thigh, which was suspected to be due to neurofibromata. In high intensity light it could be seen that there were multiple small calcifications in the soft-tissue tumour mass on the right side of the thigh. (Calcifications can occur in lipomata but are not usually seen in neurofibromata.) There was an effect of extrinsic pressure on the cortex of the upper end of the femur.

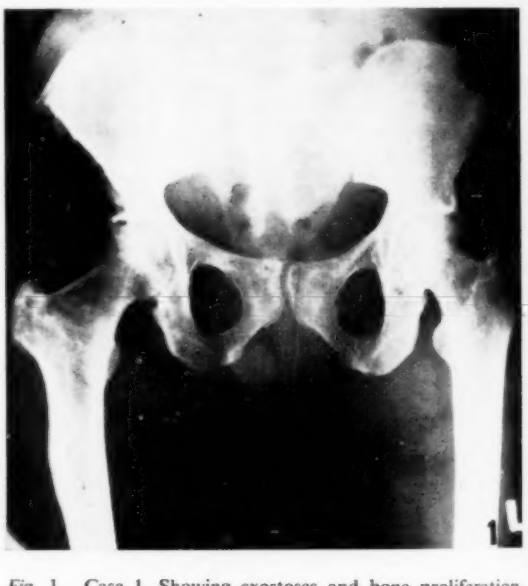


Fig. 1. Case 1. Showing exostoses and bone proliferation of right ilium, with flattening of lateral aspect.

The patient died on 29 April 1954.

Case 2

A Bantu female aged 17 years, admitted on 9 August 1953 complaining of swelling on outer border of left foot. This commenced 2 years ago and was operated on at another hospital and recurred within 2 months. From the time of origin of the swelling

the bony architecture of the foot had gradually collapsed and walking was at times extremely painful. A similar swelling removed from over the left clavicle did not recur, but a keloid developed in the scar. A 3rd swelling developed near the external meatus of the right ear 4 months back and was gradually increasing in size.

Examination. A soft swelling occupied the right external auditory meatus and appeared to be attached to both sides of the external auditory canal. There was also a soft ill-defined swelling over the antero-lateral aspect of the left foot (4 × 3 inches) with an operation scar over the swelling. Crepitus could be elicited at the ankle joint.

Diagnosis: Multiple lipomata, neurofibromata, synovioma.

X-ray. There were cyst-like areas in the bases of the 3rd, 4th and 5th metatarsals and in the tarsal bones. The bones affected showed some alteration in size and outline as compared with the bones of the right foot. Some of the intertarsal joint-spaces were enlarged,

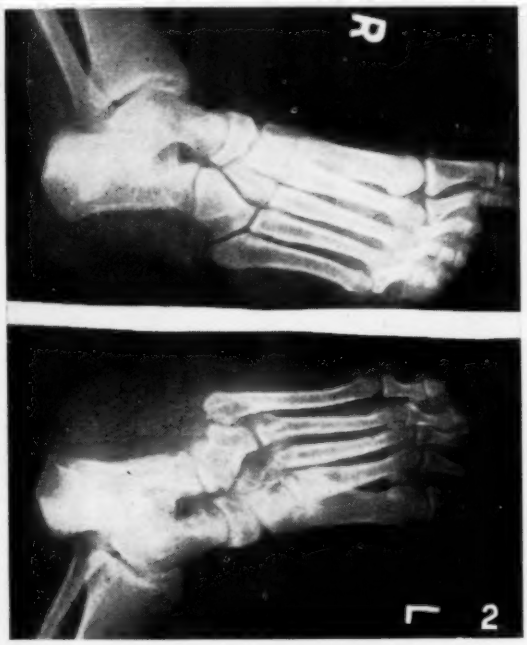


Fig. 2. Case 2. Showing deformity of tarsal bones.

best seen in the oblique views. There was associated soft-tissue swelling on the lateral aspect of the foot, with a marked varus deformity and some pes-cavus deformity. The appearances were considered to be due to neurofibromatosis.

Biopsy of meatal tumour: Plexiform neurofibroma (Dr. E. Roach).

Complete removal of the meatal tumour was found to be impossible.

Case 3

An Indian male aged 7 years, admitted on 27 February 1948 with a diagnosis of acute osteomyelitis of his right foot. This apparently developed after injury 7 days previously.

Examination. Child was in poor condition with a diffuse swelling of the right leg and foot. It was extremely tender, with medial fluctuation. Temperature 99°F. Other systems normal.

Operation. Incision along medial aspect lower leg; much pus drained.

X-Ray reports at this time were consistent with an osteitis, and a review of serial reports suggests that radiographically the response was rather tardy. Penicillin and sulphadiazine were administered. Unfortunately these films have been discarded and are not available for review.

Subsequent healing took place.

Readmission. The case was readmitted on 21 July 1953 for a 'lump' on the back, but this appeared to be due to extreme scoliosis, the curvature being to the right in the thoracic region. On examination the right glutei appeared to be weak; apart from general wasting no other abnormality demonstrated. Diagnosis was made of old poliomyelitis.

X-Ray. The spine showed gross lumbo-dorsal scoliosis, with atrophy and variation of the normal trabecular pattern of the bodies in this region. The lateral view indicated that there was anterior bulging of the posterior vertebral margins of the lower dorsal vertebrae, with resultant widening of the spinal canal and intervertebral foramina. The transverse processes of the bodies in this

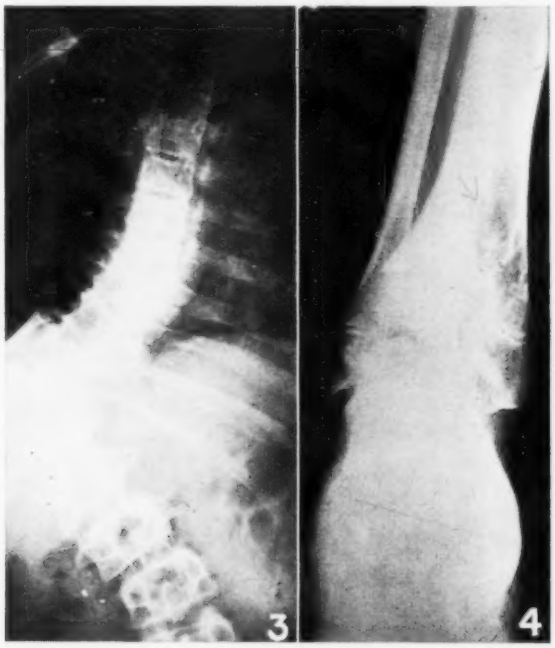


Fig. 3. Case 3. Showing gross lumbo-dorsal scoliosis with atrophy of transverse processes and vertebral extremity of 12th rib.

Fig. 4. Case 3. Showing old osteitis with cystic change.

region appeared to be atrophic and this was well seen in the antero-posterior view. The 12th rib showed atrophic thinning of its vertebral extremity. The appearance was considered to be highly suggestive of neurofibromatosis of the spine. Skeletal survey demonstrated a cystic oval area of the distal and medial aspect of the right tibia showing coarsening of trabeculation. Sclerosis on the lateral aspect had developed, and deformity of growth was that of a valgus deformity. The appearance was thought to represent a neurofibromatous lesion.

The chest film revealed no pulmonary abnormality. The right cardio-phrenic angle was occupied by the convexity of the scoliotic spine.

At the time of this X-ray report it was not known that the patient had been previously admitted for osteitis of the right tibia. When this information became available the tibial lesion was reviewed. It was then thought to represent an old osteitis with cystic change. This particular cystic type of end-result was, however, felt to be unusual, and it was subsequently suggested to us [14] that the lesion was due to infection in a neurofibromatosis deposit following trauma. Re-examination of the patient revealed numerous plexiform subcutaneous neurofibromata associated with *café au lait* skin pigmentation.

Case 4

A Native male aged 18 years, admitted on 20 August 1954 complaining of gross swelling of the right leg from early childhood. The patient stated that he was born with his 4th and 5th toes deformed, and from early childhood his right foot became progressively bigger. It was not painful except for an area over the heel and he said he could walk well enough.

Examination. Gross enlargement of right lower leg, firm and non-tender; does not pit on pressure. There is a fluctuant area over the heel. No increase of skin temperature as compared with other limb. No bruit. Femoral artery palpable. There are distended superficial veins on thigh. There is also a large soft swelling 6 × 3 inches on right buttock, superficial and non-tender. There are similar but smaller swellings all over the trunk. On questioning the patient said these had been present since his leg started swelling. Bilateral gynaecomastia present. Pus was aspirated from one of the leg swellings.

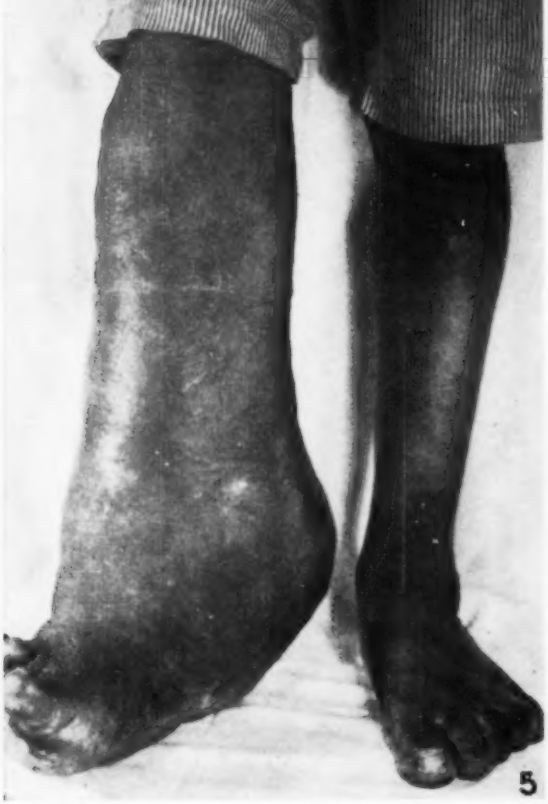


Fig. 5. Case 4. Showing gross swelling of right lower leg.

Nothing abnormal found in the *central nervous system*, the *cardio-vascular system*, or the *chest*.

Diagnosis: Neurofibromatosis, lipomatosis, phlebectasia.

X-Ray. There was a massive soft-tissue swelling in the lower half of the right leg, extending posteriorly and more medially than laterally. The tibia and fibula were widely separated. There was some localized cortical thickening on the posterior aspect of the middle third of the cortex of the tibia. The bones of the right foot were decalcified and showed areas of bone defect, particularly the

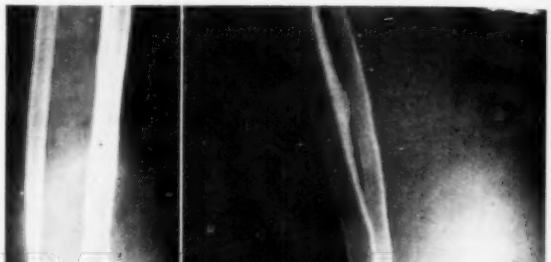
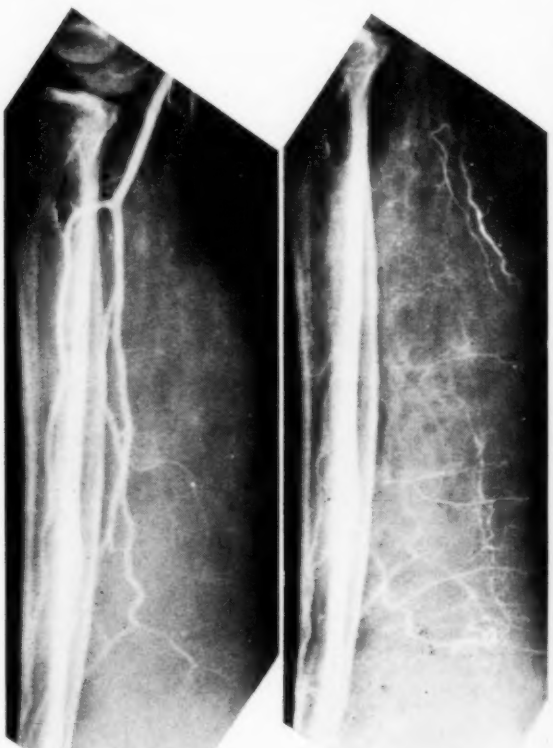


Fig. 6. Case 4. Showing separation of tibia and fibula.

Fig. 7. Case 4. Showing localized cortical thickening on the posterior aspect of tibia and the gross alteration in the bones of the foot.



Figs. 8 *and* 9. Case 4. Arteriograms.

os calcis and the cuboid. In the upper end of the right tibia there was an elongated oval area of bone absorption. A review of the skeleton did not show any other obvious changes except that a small transradiant area was seen in the upper end of the descending ramus of the ischium on the right side. The pelvis was seen to be deformed and the right femur externally rotated. There appeared to be an old healed fracture of the horizontal ramus of the left pubis. There was lumbar scoliosis convexity to the right. No well-defined transradiant areas were seen in the soft-tissue tumour mass in the right buttock.

The radiologist favoured a diagnosis of neurofibromatosis on account of the bone changes in the foot but did not exclude the possibility of lipomatosis. To exclude a vascular tumour he suggested that an arteriogram should be done. The possibility of a Kaposi sarcoma was considered.

A *Right Femoral Arteriogram* (31 August) revealed a reduced rate of flow, splaying of the vessels in the lower third of the calf, and a fine network of abnormal vessels at this level. It was found subsequently that the splaying and abnormal vessels were at the site of a haemorrhagic tumour, with sarcomatous change.

Skin Biopsy (7 September) revealed an ill-defined fibromatosis nodule, with a few hypertrophied nerve bundles as seen in a plexiform neuroma (Dr. J. Wainwright).

Operation by Mr. Stafford Meyer (23 September). Mid-thigh amputation. Evidence of gross neurofibromatosis of superficial nerves was present. The sciatic nerve was grossly thickened (1 inch thick); it was ligatured and cut.

Detailed Appearance and Histology of the Specimen (Dr. J. Wainwright).

Elephantiasis Neurofibromatosa. There is gross enlargement of the amputated limb due to a diffuse plexiform type of neurofibromatosis of the soft tissues. The sciatic nerve is grossly thickened and nodular at the site of amputation and other nerves show similar distortion.

At the ankle there is a large spheroidal haemorrhagic tumour which shows sarcomatous change. A similar smaller sarcomatous nodule is seen below the knee joint. These sarcomatous tumours appear to have grown expansively and show no involvement of bone. The bones, particularly the tarsal bones, show marked atrophy of the trabeculae, probably due to disuse.

There is infiltration of the cortex of the bones by the neurofibromatous tissue resulting in lacunar absorption of the compact bone, and widening of the Haversian canals.

In places there has been deposition of new-woven bone on the eroded bone surface. Fibrils of the tumour tissue appear to be incorporated in this new-woven bone.

The sections clearly indicate infiltration of the bones by the neurofibromatous tissue from without, and show no evidence of primary neurofibromatosis of the bones.

Case 5

A Native male aged 15 years, referred from Port Shepstone Hospital with diagnosis of spondylolisthesis L5, and admitted on 26 April 1950.

Examination. Abdomen distended owing to lumbar lordosis. No abdominal masses. No rigidity. Generally—a diffuse neurofibromatosis.

X-Ray Report (Dr. F. Gillwald, 3 May). (1) There is a gross spondylolisthesis between L5 and S1. (2) The bodies of L3, 4, 5 and S1 show considerable irregularity of density, with posterior depressions and widening of the intervertebral foramina. The

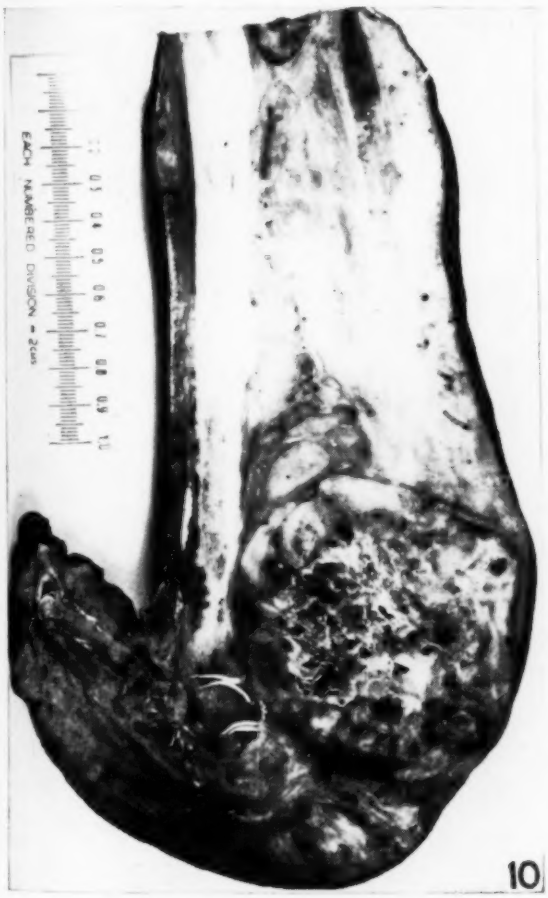


Fig. 10. Case 4. Vertical section of amputated leg showing neurofibromatous tumour and the large area of sarcomatous degeneration. The destruction of the tarsus is demonstrated.

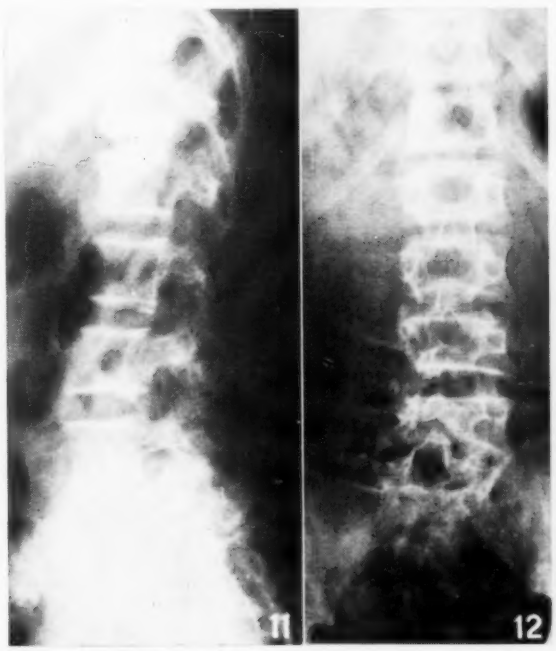


Fig. 11. Case 5. Showing spondylolisthesis between L5 and S1 and posterior depressions of vertebral bodies and widening of intervertebral foramina.

Fig. 12. Case 5. Showing erosion of pedicles on right of L4 and L5.

pedicles on the right of L4 and L5 appear to be eroded and there is a scoliosis and rotation of the vertebrae at this level. The disc spaces are intact. The appearances are those of a neurofibromatosis.

Operation (11 May). Through antero-lateral 6-inch incision, a 3-inch tibial graft taken from right tibia and wound closed. Approach through right lower paramedian incision and pelvic peritoneum opened for 2 inches over the body of S1, just to the right of the mid-line; with small chisel lower surface of L5 traumatized and hole made obliquely into body of S1. Tibial graft down into S1 against inferior surface of L5. Pelvic peritoneum sutured and abdominal wound closed.

Case 6

A Native female aged 57 years, admitted on 7 December 1954 complaining of a large growth arising from the right ankle. The patient stated that this had been there since birth, but had increased in size until it had reached the stage of interfering with her walking. Numerous smaller lumps covered the body.

Examination. The body swellings were soft and were not attached to deeper tissues. The growth surrounding the right ankle appeared to be formed by markedly thickened skin encasing numerous

Fig. 13. Case 6. Antero-posterior view of tumour.

lobulated tumours. It extended down to the ground tending to envelop the foot, and when viewed from the rear, closely resembled the leg and foot of an elephant. General examination of the patient revealed no other abnormality. The diagnosis was that of generalized neurofibromatosis.

X-Ray Report. There is a gigantic soft-tissue tumour of the lower half of the leg, more extensive posteriorly than anteriorly. The tarsal bones are deformed, and the talus and os calcis show areas of bone defect and apparent erosion. The remaining tarsal bones show a distortion of trabecular pattern. The appearances are consistent with the clinical diagnosis of neurofibromatosis.

A review of the skeleton does not reveal any other bone lesions.

A femoral arteriogram reveals that the tumour is extremely vascular, with marked displacement of the vessels, particularly of the posterior tibial artery. There is no appreciable delay in clearing of the dye considering the bulk of the tumour. There are no localized areas of stasis or pooling of dye to suggest malignant change. The venous channels are also dilated.

Operation (Mr. A. Beiles). The patient refused amputation, but agreed to an attempt at removal of the main bulk of the ankle tumour. An incision was made at the lateral aspect of the lower leg, and the posterior tibial vessels were ligated. Skin flaps were made and the tumour was dissected from above. It was extremely difficult to define the anatomy, but as much of the tumour as possible was removed.

Histology. Dr. J. Wainwright diagnosed Elephantiasis Neurofibromatosa. He reported diffuse neurofibromatosis involving collagenous tissues, and no evidence of malignancy.

DISCUSSION

1. In regard to the characteristic deformity of bone and the change in the trabecular pattern, review of the litera-

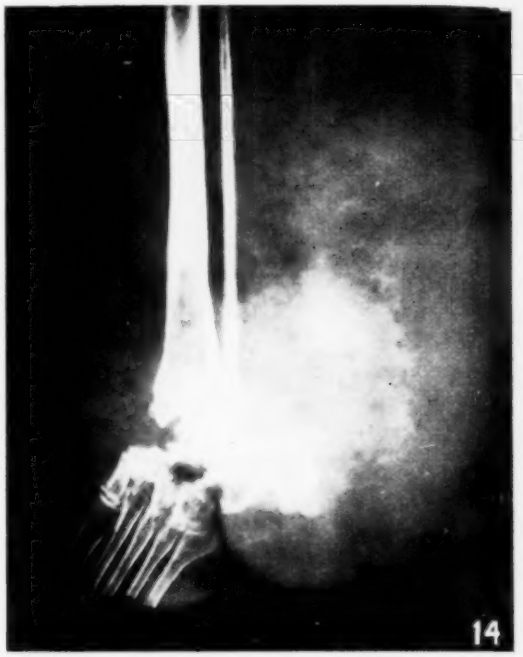


Fig. 14. Case 6. Showing tumour and bone changes in tarsus.

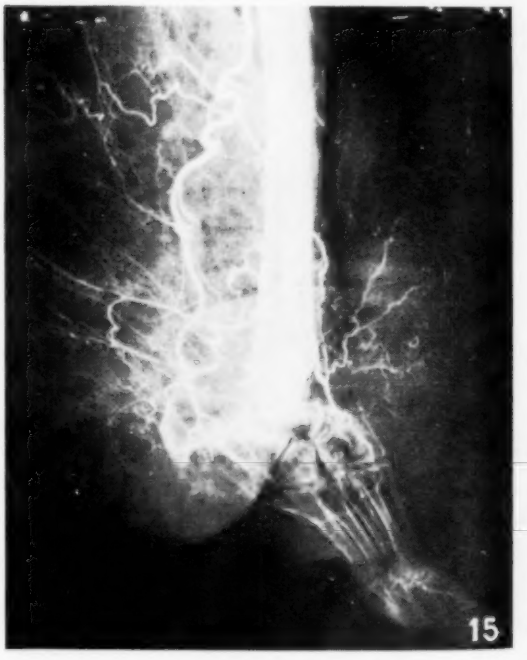


Fig. 15. Case 6. First phase of arteriogram showing increased vascularity and displacement of vessels.

ture and the study of our cases appear to indicate

(*a*) That the deformity is due to a large extent to extrinsic pressure.

(*b*) That there is infiltration of the bones by the neurofibromatous tissue from without. An excellent opportunity of studying the histology presented itself in case 4. This failed to reveal any evidence of a primary neurofibromatosis of the bones. The unusual pattern of vertebral structure in case 3 is in our opinion peculiar to bone associated with neurofibromatous lesions. It is felt that the contributing factors are:

(i) The gross disturbance of normal growth by the adjacent developing neurofibromata.

(ii) Actual invasion of bone.

There does not appear to be a satisfactory explanation of the reactive bone formation seen in the pelvis in case 1. It may be an irritative phenomenon. There is no proof of the etiology of the so-called intra-osseous bone cysts, but it may be that they appear to be intra-osseous because of proliferative bone reaction after invasion from without.

2. Scoliosis apparently occurs as an associated congenital defect, and its frequency has been mentioned in our review of the literature. It appears that it may be present without the curious twisted, almost fragile, deformity of the vertebrae found together with a gross lumbo-dorsal scoliosis in case 3.

3. Spondylolisthesis is apparently a spinal change rarely reported. In case 5 it was the main presenting feature and the reason for referring the patient to our hospital for orthopaedic control of the condition. It was only after radiological investigation that the associated neurofibromatosis was recognized.

4. We have not found any references in the literature to arteriography in this disease. In case 4 (neurofibromatous elephantiasis) are found splaying of the vessels, reduced rate of flow, residual abnormal-looking vessels, at a level which was subsequently found to be the site of sarcomatous change in a haemorrhagic tumour.

5. An impression is gained that the incidence of this disease is relatively high in the Bantu of Natal. Only 1 of the 6 cases reported was an Indian. One of us (M.F.), working for a number of years in Transvaal non-European hospitals, can recall only one case of bone changes in neurofibromatosis.

We are indebted to Dr. S. Disler, Superintendent of King Edward VIII Hospital, for permission to submit this paper, to the surgeons and physicians for their valuable co-operation, to Dr. J. Wainwright for the histology and photographs of specimen in case 4, and to Miss M. McLaggen, Department of Medical Photography, Wentworth Hospital, for the X-ray reproductions and case photographs.

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KONGENITALE ICHTHIOSIFORME ERITRODERMIE

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Kongenitale ichthiosiforme eritrodermie is 'n seldsame huidsiekte wat, sover ons bekend, nog nie vantevore by die Bantoe in Suid-Afrika beskryf is nie. Drie van die vier gevalle wat hier beskryf word, is in Bantoekinders waargeneem.

Soos die naam aandui, is hierdie siekte gekenmerk deur rooiheid van die vel met hiperkeratose. Die siektebeeld is in 1902 deur Brocq beskryf en afgesonder van die ander vorms van ichthiosis. Die eritrodermie kan reeds by die geboorte aanwesig wees, maar kan ook later optree. Die hiperkeratose kan bestaan uit 'n fyn skilfering wat skarlatiniform kan wees of dit kan in groot velle voorkom. Die afskilferende lae is deurskynend en lyk baie soos kollodion wat op die liggaam geplak is. Soms ontstaan groot barste met erosies. Die afwyking is hoofsaaklik aan die streksye en in die liggaamsplooie gelokaliseerd. Die handpalms, voetsole, gesig en kopvel is ook dikwels aangetas. Ektropion, oor-, tand- en mondmisvorminge kan voorkom. Gewoonlik is daar weinig of geen subjektiewe klagtes nie. Soos by ander genodermatoses kom blaarvorming dikwels voor. Die naels is soms verdik met subunguale hipertrofie. Hiperhidrose kan soms 'n baie lastige simptoom wees. Ook die hare toon in sommige gevalle vermeerderde groei.

Konsanguiniteit, asook sifilis van die ouers, is dikwels beskryf (Ingman,[1] Barker en Sachs[2]). Barker en Sachs[2]

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† Hoof: Dr. J. Marshall.

beskryf 'n geval van 'n 7-jarige meisie waar die vader gely het aan 'n liniëre ichthyosis hystrix. Die aandoening is meestal dominant erflik.

Dit is belangrik om kongenitale ichthiosiforme eritrodermie van ichthyosis vulgaris te onderskei wat klinies sowel as histopatologies totaal verskillend is. By ichthyosis vulgaris is die distribusie hoofsaaklik ook aan die streksye van die ekstremiteite, terwyl die buigsye, handpalms, voetsole en gesig vry bly. Die toestand ontwikkel meestal eers enkele jare na geboorte. Ook wat erflikheid betref, verskil hierdie twee siektes van mekaar daar ichthyosis vulgaris ressessief erflik is terwyl ichthyosis congenita dominant erflik is.

BESPREKING VAN GEVALLE

Gedurende 1954 was daar 4 pasiëntjies, wat met die so pas beskrewe siektebeeld ooreenstem, in die kindersale van die Algemene Hospitaal, Pretoria, onder prof. J. G. A. Davel, opgeneem. Hul siektebeelde word nou kortliks weergegee:

Fig. 1. Geval 1. Blanke kind 1 week oud met uitgebreide afskilfering van die vel.

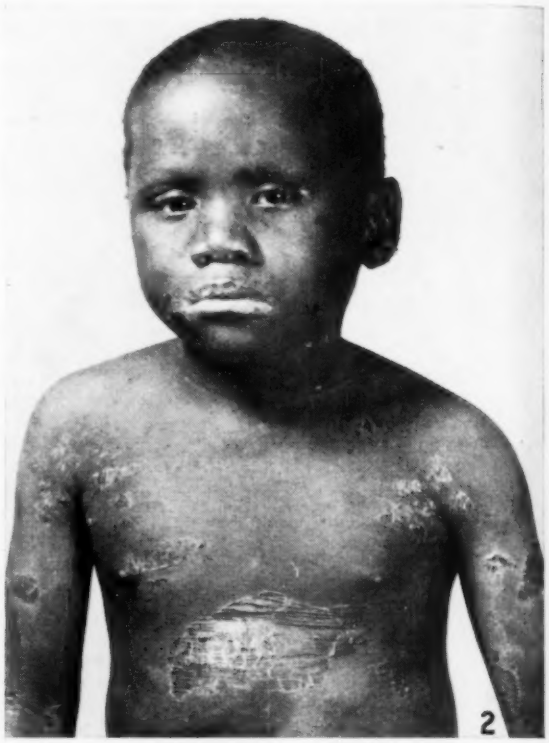

Fig. 2. Geval 2. Gesig en bolyf van 7-jarige Bantoemeidjie toon kollerige afskilfering met aantasting van die mond.

Geval 1. Baba L was 2 dae na geboorte met die klagte dat sy vel in groot stukke afkom, opgeneem. Dit was met die bevalling alreeds duidelik te sien. In die familiegeskiedenis was daar niks van belang nie en 'n dergelike veltoestand was by geen ander familielid bekend nie. Die baba was die 3de kind, die 1ste 'n dogter van 3 jaar, die 2de 'n seuntjie van 17 maande. Beide laasgenoemde was heeltemal gesond. Die moeder was gesond gedurende swangerskap en die bevalling het normaal verloop. Die geboortegewig was 7 lb.

Met ondersoek was die voorkoms van die baba soos in Fig. 1 aangetoon. Hy was baie siek en die mees opvallende afwyking was dié van sy vel. Daar was 'n besondere rooi egalige eriteem oor sy hele liggaam versprei. Groot gedeeltes van sy vel het afgeskilfer. die skilfers was van 'n lig goudkleurige voorkoms en het die duidelike indruk geskep asof hierdie baba in kollodion gedompel was. Die velbarste was die mees uitgesproke in die huidplooie, oor die buik, bors en rug (Fig. 1). Die streke, waar vervelling alreeds plaasgevind het, was besonder rooi; daarop het direk al weer fyn barsies verskyn en dit was blykbaar pynlik met aanraking. Op die kopvel was hierdie toestand ook teenwoordig met slegs geringe afskilfering. 'n Konjunktivitis was in beide oë teenwoordig en beide ooglede het verdik voorgekom met 'n geringe graad van ektropion. Die ore het albei 'n matige misvorming van die skulp (helix en antehelix) vertoon. Daar was geen mond- of anusmisvorming nie. Die naels was ook normaal.

Die pasiëntjie was in 'n broeikas geplaas met 'n vogtigheidsgraad van 80-90 en sy vel was met olyfolie behandel. Gedurende die eerste week was penisillien toegedien en 'n voeding van verdunde aangesuurde afgeroomde koeimelk voorgeskryf, aangesien sy

Fig. 3. Geval 2. Onderste ekstremiteite met uitgebreide barste en afskilfering.

moeder geen borsmelk gehad het nie. Kortisoon, 12·5 mg. elke 6 uur, was dadelik toegedien. Na 6 dae was die kortisoon met cortrophin Z, 10 mg. elke 2de dag, vervang. Kaliumkloried, 7½ gr. t.i.d., was saam gegee.

Aanvanklik het hierdie pasiëntjie baie verbeter en daar was periodes van so lank as 3 dae waar geen vervelling plaasgevind het nie, maar die proses het weer opnuut begin en geleidelik erger geword. Hoë dosisse van vitamin A was by die behandeling bygevoeg, sonder enige verbetering. Die 14de dag na opname het hy begin braak en 'n erge diarree ontwikkel. Alle terapie was gestaak en Hartmann se oplossing per mond toegedien. Vir 'n meegaande koors van 102°F was geen parenterale oorsaak te vinde nie, derhalwe was hierdie toestand as 'n infektiewe gastro-enteritis beskou. Daar die baba vinnig agteruit gegaan het, moes 'n insnyding vir parenterale vogtoediening gedoen word; ten spyte daarvan egter is hy die 21ste dag oorlede. Toestemming vir 'n lykskouing was geweier. Urine-ondersoek en bloedtelling het geen afwykings getoon nie. Die bloed Kolmer was negatief.

Geval 2. 'n Bantoemeidjie van 7 jaar was vanaf 'n plattelandse hospitaal, met die diagnose van pellagra, verwys. Geen familielid het saamgekom nie en geen behoorlike geskiedenis kon verkry word nie. Sy self het beweer dat so lank as wat sy kon onthou, haar vel in dié toestand was. Ondersoek het geen noemenswaardige afwykings openbaar nie behalwe die opvallende velafwyking. Die hele liggaam, maar veral die ekstremiteite, het kolle van verskillende grootte getoon waar die vel besig was om af te skilfer. Die vel in die huidplooie was verdik en het lichenifikasie vertoon (Fig. 2 en 3). Die voorkoms was byna soos die van 'n opgedroogde plaasdam. Opvallend was ook die teenwoordigheid van enkele klein bullae, veral op die gesig. Die lippe het ook gedurige afskilfering vertoon met barsies in die mondhoeke waarin voortdurend ligte infeksie voorgekom het.

Die enigste behandeling wat hierdie pasiëntjie gekry het, was vitamin A 20,000 E. per dag, maar ook hier was geen voordelige effek opgemerk nie. Hierdie veltoestand het afwisselend verbeter en weer versleg en so is die pasiëntjie uiteindelik huis toe. 'n Velbiopsie van so 'n aangetasde area is deur dr. P. J. Barnard van die Instituut van Patologie, Pretoria, ondersoek en sy verslag volg: „Huidsnitte toon kollerige verspreide streke van hiperkeratose en parakeratose, beide saam en/of apart met afskilfering as gevolg. Verskeie sweetklieruitmondings skyn verstop te wees. Die granulêre laag is deurgaans effens verwyd en die suprapapillêre retelaag het òf verdun òf heeltemal verdwyn. Basaalselle bevat hier en daar helder vakuoli en vertoon meer mitotise aktiwiteit as normaal. Geen epidermolise is aangetref nie. In die buitenste derde van die

Fig. 4. Geval 3. Bantoebaba 1 week oud met ligte afskilfering van die vel, veral oor die buik.

korium is menige uitgesette kapillêre vate (òf bloedvate òf limfvate wat nie met sekerheid vasgestel kan word nie), sowel as selophopings, meesal limfositêr, teenwoordig. Geen plasmaselle en eosinofielselle is gevind nie. Die elastika het herdwyse effens opgebreek soos by enige chroniese ontstekingsproses van die dermis verwag mag word. Huidaanhangsels kom normaal voor. Hierdie afwykings pas goed in by die hipertrofiese stadium van erythrodermia ichthyoformis congenita.'

Geval 3. Hierdie Bantoekind was in die kraamafdeling van die Pretoriase Algemene Hospitaal gebore. Daar was direk na geboorte opgemerk dat sy vel 'n fyn skilferende voorkoms gehad het. Dit was nie baie erg nie en byna skarlatiniform in voorkoms (Fig. 4). Dit het afgeskilfer en weer 'n lagie gekry wat na enkele dae weer begin afskilfer het. Die distribusie van die aandoening was dieselfde as in die vorige gevalle, hoewel van geringer graad as in die eerste geval. Na ontslag was die baba tot dusver nog nie gesien nie.

Fig. 5. Geval 4. Bantoebaba 8 maande oud toon droë skilferende huid met barsies.

Geval 4. 'n Bantoebaba van 8 maande was opgeneem in 'n toestand van dehidrasie met aanval van gastroenteritis van 'n week se duur. Met ondersoek was opgemerk dat die vel droog was met fyn barsies en afskilfering oor die hele liggaam (Fig. 5). Volgens die moeder was die kind se vel by geboorte rooi van kleur en soos sy dit uitgedruk het, „die kind was met masels gebore". Hierdie aanvanklike eriteem het in 'n afskilfering na enkele dae oorgegaan en die hele proses is voortdurend tot sy dood toe herhaal. Die letsels was hier ook hoofsaaklik beperk tot die ekstremiteite, huidplooie, buik en rug. Die mond, ore en oë het geen afwykings getoon nie.

BESPREKING

By al 4 gevalle so pas beskryf, was hierdie siekte waarskynlik by geboorte teenwoordig. Alleen in geval 2 bestaan daar twyfel hieromtrent omdat 'n bevredigende geskiedenis nie verkry kon word nie. Die eritrodermie by geboorte is in 2 van die gevalle self waargeneem en die beskrywing van die moeder van geval 4 as sou die baba se vel soos die van masels by geboorte gelyk het, is baie treffend. In 3 van hierdie gevalle het ook die handpalms op die tipiese manier afgeskilfer en die lokalisasie van die velaantasting is soos dié van beskrywing van die tipiese siektebeeld; daarby die ongeneesbaarheid, selfs met kortisoon en ACTH, en die herhaling van die hele proses, is tot die besluit gekom dat hierdie 4 gevalle tot die siektebeeld van kongenitale ichthiosiforme eritrodermie behoort.

Daar bestaan verskil van mening oor die selfstandigheid van hierdie siektebeeld. Volgens meeste ondersoekers is dit alleen 'n ligte vorm van kongenitale ichthiosis. Die oudste beskrywing van hierdie siekte is in 1792 deur Richter en in 1828 deur Steinhauser[3] gegee onder die naam ‚De Singulari Epidermides Deformitate'. In 1880 het Perez[3] die siekte beskryf onder die titel ‚Sclérose générale de la peau chez un nouveauné'. Daarna is die siekte herhaalde kere onder meer as 20 verskillende name beskryf. Die poging om 'n vergelyking te tref met natuurvoorwerpe blyk uit die volgende benamings: ‚Alligator-boy', ‚cutis testacea', ‚harlequin foetus', ‚desquamation collodionnée', ‚fish-skin disease'.

Uit die beskrywings van Hebra[4] en Kaposi[5], blyk dit dat hulle hierdie toestand nie onderskei het nie van die seborrhoea squamosa van pasgeborenes, wat 'n verbygaande siekte is.

Riecke[6] het ichthyosis congenita in 3 tipes verdeel:

1. Ichthyosis congenita gravis. Hierdie kinders word altyd te vroeg gebore. By die geboorte is hulle omhul met 'n harde hiperkeratotiese massa met diep barste daarin—die sogenaamde harlekyn fetus. Kongenitale misvorminge van oë, ore, mond en anus is uitgesproke.

2. Ichthyosis congenita larvata. Die kinders word gewoonlik voltyds gebore maar leef meestal net enkele ure of 'n paar dae. Daar is selfs gevalle beskryf wat in lewe gebly het. Die kongenitale afwykings is minder uitgesproke. By beide bogenoemde gevalle ontstaan die afwyking op ongeveer die 4de of 5de swangerskapsmaand.

3. Ichthyosis congenita tarda. Hierdie siektebeeld kom ooreen met die kongenitale ichthiosiforme eritodermie soos deur Brocq beskryf. By die geboorte is dikwels geen afwykings te bespeur nie of hoogstens 'n

diffuse rooiheid van die huid. Kongenitale afwykings is òf baie gering òf ontbreek heeltemal.

Ingman[1] het 3 kinders uit een gesin beskryf: die 1ste was 'n 7 maande doodgebore kind, die 2de het net een maand oud geword, en die 3de het in lewe gebly. Hy haal ander gevalle aan waar miskrame voorgekom het voordat ichthiotiese kinders gebore is. Verder gee hierdie skrywer 'n uitvoerige beskrywing van 111 bewysde en 34 twyfelagtige gevalle, wat hy uit die literatuur versamel het. Hy toon aan dat die horinglaag in sy gevalle verhoogde gehaltes cholesterien en vetsuur esters bevat. Volgens hom sou dit die oorsaak wees van die abnormale verklewing van die horinglae wat die rede is waarom dit nie afgestoot word nie.

'n Vermoedelike variant van hierdie siekte is in 1924 deur Mendes da Costa[7] onder die naam erythro- et kerato-dermia variabilis beskryf. In sy proefskrif bespreek Noordhoek[8] hierdie siekte breedvoerig met vermelding van enkele eie gevalle. Die veranderlikheid van die areas van eriteem en ook soms van dié van hiperkeratose, tree steeds op die voorgrond. Twee van sy gevalle, 'n broer en suster, sou volgens hom ook inpas by die diagnose van kongenitale ichthiosiforme eritrodermie.

Wat die histologiese beeld betref is deur enkele ondersoekers[9, 10] onlangs die aandag gevestig op die onvolledige akantolise wat in die stratum mucosum van die bulleuse vorms voorkom. Die stratum granulosum is meestal aanwesig, maar kan soms ten dele ontbreek. Hiperkeratose is sterk uitgesproke met onreëlmatige areas van parakeratose. Lapière[10] het ook diskeratosis met *corps ronds* waargeneem. Marshall en Martin[9] bespreek breedvoerig die histologiese beeld van 'n geval en wys op die ooreenkoms met benigne kongenitale pemfigus. Lapière vestig die aandag op die ooreenkoms met die bulleuse siekte van Darier. Laasgenoemde 2 siektes word deur sommige skrywers as dieselfde siektebeeld beskou. Marshall, Martin[9] en Lapière[10] verdedig egter die standpunt dat nieteenstaande die duidelike histologiese ooreenkomste, dit aparte siektes is. Daar bestaan geen gronde nie vir die mening deur sommige geuit dat epidermolisis bullosa ook 'n variant van kongenitale ichthiosiforme eritrodermie is, aangesien die histologiese beeld ook verskil. Die bullae by die epidermolysis bullosa is altyd sub-epidermaal en nooit intra-epidermaal geleë nie soos dit die geval by kongenitale ichthiosiforme eritrodermie is. By laasgenoemde is ook nooit 'n porfirinurie gevind nie, wat vermoedelik wel altyd die geval is by epidermolisis bullosa[9].

Diagnostiese moeilikhede mag ondervind word met die siektes van Ritter von Rittershain (dermatitis exfoliativa neonatorum) en Leiner (eritrodermia desquamativa neonatorum). Die eerste is vermoedelik 'n piogene infeksie van pasgeborenes en tree meestal tussen 2 en 5 dae na geboorte op. Groot areas van die epidermis kan afdop (eksfolieer) om groot rou sere te laat. Met moderne behandeling genees die siekte volkome. Die siekte van Leiner tree meestal later op na die 20ste dag. Dit kan in die begin baie op 'n seborrhoeiese of infantiele ekseem lyk. Die siekte is aanvanklik hoofsaaklik in borsgevoede babas waargeneem en het dikwels 'n dodelike verloop. Cole *et al.*[11] het in 'n onlangse artikel die mening uitgespreek dat hierdie siekte dieselfde is as Ritter se siekte, alleen in kinders met 'n hoër weerstand teen die infeksie. Albei siektes is te onderskei van kongenitale ichthiosiforme eritrodermie deurdat dit onder moderne behandeling totaal genees.

'n Ander siekte wat onderskei moet word van die groep van ichthyosis congenita is die s.g. seborrhoea squamosum neonatorum waarby die oppervlakkige lae van die epidermis in groot velle afgestoot word. Dit is 'n verbygaande toestand en die geval deur Finlay en Baund[12] beskryf behoort waarskynlik tot hierdie groep. Die toestand berus vermoedelik op 'n vermeerderde afskeiding van vernix caseosa wat nie vinnig genoeg afgestoot word nie.

SLOTSOM

Die skrywers stem saam met Layman en Murphy[13] dat kongenitale ichthiosiforme eritrodermie die ligste vorm van kongenitale ichthiosis is en met die ichthiosis congenita tarda van Riecke ooreen stem.

OPSOMMING

Vier pasiëntjies, 2 pasgeborenes en die ander 8 maande en 5 jaar onderskeidelik word beskryf met die toestand kongenitale ichthiosiforme eritrodermie. Hierdie siekte is gewoonlik by geboorte teenwoordig en bestaan uit 'n voortdurende afskilfering van die vel. Dit kan van 'n geringe of baie erg graad wees en is hoofsaaklik tot die liggaamsplooie en strekoppervlaktes beperk. 'n Oorsig van die literatuur toon dat hierdie velafwyking alreeds in 1792 beskryf is en hierna weer herhaaldelik onder omtrent 20 verskillende benamings. 'n Mate van verwarring heers oor die regte klassifikasie en die skrywers reken dat hierdie toestand 'n ligter vorm van kongenitale ichthiosis is en met die ichthyosis congenita tarda van Riecke ooreenstem.

SUMMARY

Four cases, 2 newborn, 1 of 8 months and 1 of 5 years, with congenital ichthyosiform erythrodermia are reported. This disease is usually present at birth and is characterized by erythrodermia and a continuous desquamation of the skin of varying degree. This may be confined to the extensor areas and the skin flexures or may involve the entire skin.

Reference is made to the literature on this subject. The condition was reported for the first time in 1792, since when it has been reported under about 20 different names. There is some disagreement as to the correct classification, but the authors submit that this disease is the mildest form of ichthyosis congenita according to Riecke's classification.

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ACUTE DILATATION OF THE STOMACH

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Acute dilatation of the stomach has long been recognized as a clinical entity, yet it is only during the last few years that its pathogenesis has come to be understood, and too often it is still attributed to mechanical obstruction, 'post-operative hypersecretion', or simply post-operative aerophagy. Though not frequently met in general practice its importance must not be underestimated. It may occur when least expected, sometimes after the most trivial of operations, and occasionally unassociated with any operation at all. It is a serious condition (Starr[1] reports a mortality of 40%) in the best hands. In a country practice, unaided by laboratory reports on the state of the plasma electrolytes, it is frankly dangerous. Only early diagnosis, a thorough understanding of its pathogenesis, and bold treatment can avert disaster.

Two cases have been encountered during the last year, differing in their development, treatment and outcome, and illustrative of several aspects of the condition.

CASE 1

Mr. S. was a healthy European male of 54 years, over 6 feet tall and weighing approximately 240 pounds. He had undergone an operation for repair of a small umbilical hernia. The anaesthetic consisted of pentothal, nitrous oxide, oxygen, ether and Flaxedil, and was administered by myself. The immediate post-operative course appeared in no way unusual, but on the 2nd day, when the patient had not passed flatus, a purgative was given. On the 3rd day there was still no passage of flatus and an enema was administered, with a negative result. By the 4th day the abdomen was somewhat distended and the patient began to vomit—at first small amounts of greenish fluid, increasing steadily in quantity throughout the day and later of a turbid, brownish colour.

When the patient was seen on the 5th post-operative day it was immediately evident that his condition was poor. He was slumped in bed, listless and apathetic, complaining of thirst. His tongue was dry and he was clearly dehydrated. The pulse rate was 88 per minute; the blood pressure 105/60 mm. Hg. The temperature was normal. His abdomen was slightly distended and bowel sounds were absent. Urinary output was diminished (800 ml. in the ensuing 24 hours) and the specific gravity high. Fantus' test for chlorides[2] was not performed as the reagents were not available. Plasma-electrolyte concentrations were not measured at any time as the results could not have been received in less than 4 days.

A diagnosis of acute dilatation of the stomach was made, and confirmed radiographically. Gastric suction, with intravenous fluid-replacement, was commenced forthwith; during the ensuing 24 hours 2½ litres of brown fluid were aspirated and 4½ litres replaced intravenously (3 litres of 5% dextrose in water, and 1½ litres of normal saline).

On the 6th day his condition had improved slightly. The blood pressure was now 115/85 mm. Hg (still lower than normal for him) and his tongue was not as dry as before. Urinary output, however, was only 700 ml. in 24 hours, and dark, foul-smelling fluid was still being aspirated from the stomach.

In spite of the diminished urinary output potassium chloride (KCl) was now added to the intravenous fluids. It was estimated at this stage that he required about 8 litres of fluid for re-hydration, about 160 mEq. of potassium and a similar amount of chloride (12 g. KCl). Neither serum nor Darrow's solution was available, and only 4 g. of KCl solution could be found in the town. This was given at the rate of 1 g. per litre of fluid.

Under the circumstances it appeared to be dangerous to give 8 litres of fluid over the next 24 hours, and this assumption was confirmed on the 7th day when, in spite of receiving only 3½ litres, he developed bilateral pulmonary oedema. Two litres of faeculent fluid were aspirated during that day. The urinary output fell to 400 ml.

In the next 24 hours he received 3 litres of fluid (including 1 litre of dextrose in saline) intravenously, but over 3½ litres of faeculent gastric fluid were aspirated. Though grossly dehydrated his lungs were very moist now, and at times he became cyanotic.

At this stage his relatives insisted on having him transferred to Cape Town by air. During the flight he lapsed into coma and died as we stood by powerless to assist him in any way.

CASE 2

Ten months later the second case was seen. On 2 August 1954 Mr. M., a European male of average build, 56 years old, developed what appeared to be a typical attack of gastro-enteritis, with pyrexia, vomiting and diarrhoea. Treatment with sulphaguanidine and a bismuth mixture brought a prompt recovery and 3 days later he was feeling quite well. That evening, however, he started vomiting again, and by the next day the vomitus was copious, dark in colour and foul-smelling. There was no sign of improvement during the day, and early the next morning he was admitted to hospital, where 900 ml. of faeculent fluid were aspirated from the stomach.

On examination he presented a listless appearance, could not concentrate on conversation and at one stage actually got out of bed with the intravenous apparatus *in situ*. He was dehydrated but his tongue was not completely dry. The pulse rate was 74 per minute; the blood pressure 125/85 mm. Hg. Temperature normal. There was slight distention of the epigastrium, but no abdominal tenderness. Auscultation elicited occasional faint propulsive bowel sounds, and 4 times during the next day a small amount of flatus was passed per rectum. The urinary output was good (1,200 ml. during the ensuing 24 hours), the specific gravity was low (1·008), and tests for albumen, sugar and ketones were negative. Fantus' test showed a chloride concentration of less than 1 g. per litre. Gastric fluid was being aspirated by Wangenstein's apparatus, and during the ensuing 24 hours totalled 4 litres. It was turbid, dark brown and faeculent, and its reaction was strongly alkaline. Here, for the same reason as in case 1, estimations of plasma electrolytes were not carried out.

Within 12 hours of admission a confident diagnosis of acute dilatation of the stomach was made. The quantity and character of the gastric fluid, the state of alkalosis and the minimal bowel sounds all pointed strongly in that direction. Radiography showed a dilated stomach and several dilated loops of small bowel with fluid levels (this in no way invalidated the diagnosis as it is known that the duodenum and jejunum are frequently involved with the stomach). Although the possibility of intestinal obstruction was borne in mind throughout, it was felt that acute potassium depletion had caused the condition and treatment was carried out on that assumption.

It was estimated from examination of the urine, the character

of the gastric fluid and the general condition of the patient, that he would require at least 7 litres of fluid during the next 24 hours, and about 12 g. of KCl intravenously. The proportion of dextrose solution to saline was to be controlled by the results of the Fantus test. A daily fluid-balance determined the fluid requirements for each ensuing period of 24 hours from 8 a.m. to 8 a.m.

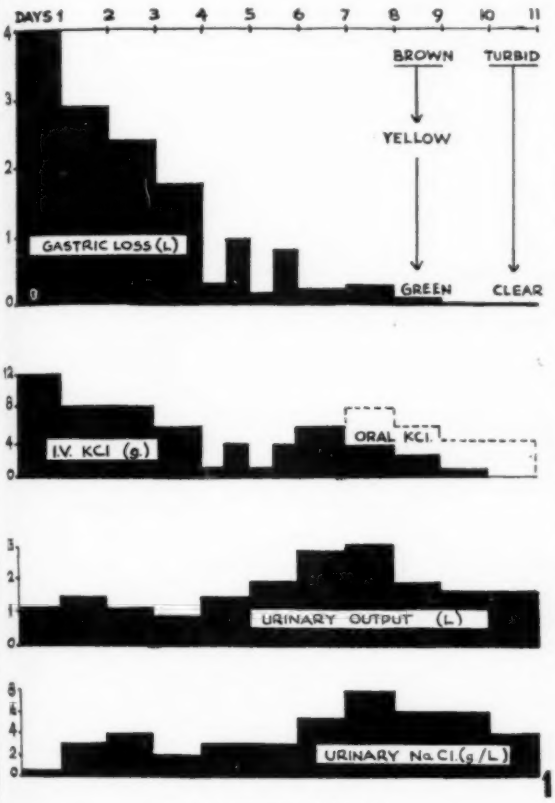

Fig. 1. Progress of case 2.

Fig. 1 shows the course during the period of treatment. Two points of particular interest appear. The first is the reaction of the stomach and bowel to KCl administration. The quantity and character of the gastric fluid rapidly returned to normal while KCl was being administered and by the 4th day of treatment less than 2 litres of clear green fluid was withdrawn in 24 hours. Bowel sounds were audible and a small soft stool was passed. At this stage it was decided to observe the response to variations in the KCl intake during the 5th and 6th days.

Starting at 8 a.m. on the 5th day the next 48 hours were divided into 4 12-hour periods during which the amount of KCl added to the intravenous fluids was to be varied. From 8 a.m. to 8 p.m. on the 5th day only 1 g. of KCl was given. During the next 12 hours 4 g. KCl were given. This was repeated on the 6th day.

The response was dramatic (Fig. 1). The sudden diminution in KCl intake was followed by an increase in gastric fluid to 1 litre in 12 hours, and a return to the brown, turbid, foul-smelling state. A rise in KCl intake during the second 12 hours was followed by an equally definite diminution in the gastric fluid to 200 ml. in 12 hours. This pattern was repeated on the next day.

By the 8th day, although the patient was now receiving most of his fluids, as well as KCl and NH_4Cl, orally, the intravenous KCl administration was still 4 g. per day, and only by the 10th day was the intravenous route abandoned.

The second point of interest is the marked and sudden diuresis accompanied by an increase in the urinary chlorides, which occurred on the 7th day, when intravenous fluids were being decreased. This event heralded recovery and by the 12th day the patient was on a normal diet again. Its significance is discussed later.

Soon after recovering the patient was sent to Cape Town, where thorough clinical, radiographic and laboratory examinations were carried out. Nothing abnormal was found. At present, after 4 months, he is still well.

ETIOLOGY

As long as 30 years ago electrolyte deficiencies were already thought to be responsible for some cases of paralytic ileus, but attention centred mainly on the sodium and chloride ions. It was not until 1943 that Ariel and his colleagues[3] reported 5 cases of post-operative ileus associated with a sodium-chloride-refractory hypochloraemia, in which recovery took place after parenteral and oral administration of protein; others had noticed an equally favourable response in cases receiving serum intravenously.[1] This led to work on other electrolytes and in 1949 Randall and others[4] suggested potassium deficiency as a cause of post-operative ileus and started using potassium in their treatment. Since then numerous reports on its value in paralytic ileus and gastric dilatation have appeared.[5–9]

At the same time experimental work has supported the clinical findings.[10–12] Of particular interest are the experiments of Streeten and Williams on NaCl-depleted dogs. Intestinal paralysis was induced only when intracellular potassium was lost as a result of adreno-cortical activity due to chloride deficiency. *In vitro* experiments on intestinal segments immersed in different electrolyte media showed that a NaCl-deficient medium could abolish intestinal movements, but a potassium-deficient medium failed to do so. In the former case a low potassium concentration was demonstrated in the bowel wall.[6]

These observations indicate that intracellular rather than extracellular potassium deficiency is responsible for intestinal paralysis, and this is not surprising, considering that nerve conduction, acetylcholine formation and cell irritability are all dependent on the potassium level.[1, 13]

The maintenance of a constantly high intracellular potassium concentration is therefore of great importance. Although cell membrane is usually permeable to them, potassium ions (K^+) do not occur in osmotic equilibrium on either side of the membrane, the intracellular K^+ concentration (144 mEq./litre of cell fluid) being far greater than the extracellular K^+ concentration (5 mEq./litre). This gradient can only be maintained by active cation transfer, the small amount of energy required being supplied by the metabolizing of glucose.[13] The tendency for sodium ions (Na^+) to enter the cell by osmosis is overcome by the same process.

The conditions under which K^+ is lost from the cell are still incompletely understood. Experimental work has shown that the cell, far from being impermeable to Na^+ ions, will under certain circumstances attract them at the expense of K^+.[13–15] For instance, in a state of K^+ deficiency a small amount of Na^+ will enter the cell. If a NaCl infusion, potassium-free, is now given even more

Na^+ enters the cell, displacing K^+. If K^+ is infused a sodium diuresis occurs.

Deoxycortone acetate, apart from its well-known action of increasing K^+ excretion in the urine, has been shown to produce a similar exchange of Na^+ for cellular K^+ in experiments on rats.[16] ACTH and cortisone also cause an increase in urinary K^+ and, clinically, adrenocortical hyperactivity has the same effect.[6, 14, 17]

Furthermore K^+ conservation in states of depletion is notoriously poor. Even during starvation the kidneys continue to excrete K^+ which is lost from the cell during protein katabolism.

These mechanisms are seen at work in the 2 cases described. In case 1 an intravenous infusion poor in potassium and containing NaCl resulted in a rapid deterioration, with salt and water retention ending in pulmonary oedema. In case 2 adequate potassium administration resulted in a return to normality with a chloride, and presumably a sodium, diuresis on the 7th day.

Turning now to the role of K^+ in the digestive system, it becomes evident that it is important not only in cell metabolism but also in maintaining the composition and reaction of the digestive juices, and the activity of their enzymes.[1] This is effected by variations in the ratio of the bulk ions Na^+, K^+ and Cl^- in the different parts of the gastro-intestinal tract. Thus K^+ and Cl^- occur maximally in the proximal or 'secretory' portion, while Na^+ (and HCO_3^-) occur maximally in the distal or 'absorptive' portion, where the K^+ level is very low. K^+ and Cl^- are presumably essential for the maintenance of function in the proximal gut, where they occur in concentrations far in excess of the plasma values (11·2 mEq. K^+ and 116·2 mEq. Cl^- per litre are average figures for 'recent ileostomy fluid'[4]).

It is precisely these differences between the proximal and the distal gut which, according to Starr, justify the recognition of acute dilatation of the stomach as an entity apart from distal paralytic ileus, though related to it. K^+ and Cl^- deficiencies are liable to affect the proximal gut sooner than the distal gut. Conversely recovery from the deficiency state occurs earlier in the distal than in the proximal gut, because of its lower K^+ and Cl^- requirements, and gastric paralysis with hypersecretion may be maximal while bowel sounds and even bowel actions occur. Furthermore NaCl administration alone may effect recovery in cases of distal ileus, but will only aggravate a proximal ileus.

That the 2 conditions will overlap is to be expected and in all probability that is what occurs in the majority of cases. Nevertheless the predominant element should be recognized and treatment guided accordingly.

In both of the cases described the stomach was predominantly involved and in case 2 intestinal peristalsis was not abolished at any time.

PATHOGENESIS

When acute dilatation of the stomach is encountered it is almost always after an operation, the maximum incidence being on the second post-operative day.

About 2½ litres of gastric juice, containing 9—116 mEq. Na^+, 0·5—32·5 mEq. K^+, and 7·8—154·5 mEq. Cl^- per litre, are normally secreted per day, and totally reabsorbed in the intestines.[4, 18] A quick calculation will show the almost unbelievable fluid and electrolyte depletion which may follow non-reabsorption of this juice over a period of 3 or 4 days. Therefore, where an operation has been preceded by diarrhoea or vomiting, and where the post-operative period has been associated with further losses of fluids and electrolytes, particularly due to prolonged gastric suction, a potassium deficiency may be expected. When, in addition, treatment consists of saline infusion without potassium replacement an even greater, rapidly progressive, hypokalaemia is inevitable.

Acute post-operative dilatation of the stomach, however, does not occur only after abdominal operations, or in association with starvation, vomiting and gastric suction. It may occur after the most trivial surgical procedure on any part of the body. The mechanisms whereby K^+ depletion arises in these cases have already been mentioned. The role of urinary K^+ loss deserves further attention.

The post-operative changes in water and electrolyte metabolism are today well known. They occur even after minor operations, where hydration is adequate and intravenous infusions unnecessary. The important features are a retention of Na^+ and water, accompanied by a marked increase in K^+ excretion in the urine lasting for 48—72 hours.[14, 17, 19] This K^+ loss may amount to 'the equivalent of 10 g. KCl' in the first 3 post-operative days,[11] resulting in a negative K balance, at least as long as food is not taken.

It has been suggested that these changes are the result of adrenocortical hyperactivity initiated by surgical trauma. This view is based on the observation that ACTH and cortisone can cause all the changes seen post-operatively, including an eosinopenia as well as increased K^+ and nitrogen excretion.

It is probably not quite as simple as this, however, and Le Quesne[17] suggests that the Na^+ and water retention, at any rate, depends on at least 3 factors: (*a*) the secretion of an antidiuretic hormone by the posterior pituitary, causing primary water retention, (2) the secretion of salt-retaining corticoids by the adrenal cortex, and (3) renal haemodynamic factors. Whatever the exact mechanism, the result may again be a serious K^+ depletion in the presence of a raised extracellular Na^+ concentration.

So much for post-operative ileus. Of equal interest are those conditions associated with hypokalaemia where no operation has been performed. These include starvation,[7] prolonged diarrhoea or vomiting, whatever the cause (particularly in pyloric stenosis, infantile diarrhoea,[20] ulcerative colitis and steatorrhoea), diabetic ketosis, and chronic nephritis. It is surprising that paralytic ileus and acute dilatation of the stomach are so seldom reported in the literature on these conditions, but the fact that they do occur[7] is important, for here one has the opportunity of studying the condition uncomplicated by metabolic changes due to operation. Thus it may be found that the simple administration of one or two electrolytes brings about a recovery, and in the case described here it was actually demonstrated that improve-

ment and deterioration in the condition closely followed the variation in the amount of KCl administered.

Even in this case, however, adrenocortical hyperactivity cannot be entirely excluded as a contributory factor. Firstly, the amount of potassium lost at the onset of diarrhoea and vomiting alone could not have been very great, and secondly, although the chloride (and sodium) diuresis on the seventh day *may* have been due to the replacement of intracellular K^+, as suggested earlier, it may equally well have been due to the cessation of adrenocortical hyperactivity. Estimations of the K^+ lost in the urine would have been of tremendous interest.

That potassium plays an important, if not the most important, role in gastro-intestinal paralysis is no longer doubted. The exact degree to which adrenocortical activity contributes is far more uncertain.

DIAGNOSIS

Prompt and efficient treatment depends on early recognition. Diagnosis involves not only the discovery of adynamic dilatation of the stomach, but an assessment of the entire state of the fluids and electrolytes.

Copious vomiting in the early post-operative period will draw immediate attention. The more insidious case will continue to escape early diagnosis as long as surgeons refuse to use their stethoscopes. Undue delay in the onset of propulsive bowel sounds should at all times arouse suspicion, and the routine use of purgatives or enemas on the second or third post-operative day is not only unnecessary but may be positively harmful where the bowel is still in an adynamic state, or where bowel movement is just starting.[21] In case 1 diagnosis and treatment were delayed for 4 days, whereas in case 2 treatment was begun even before a definite diagnosis had been reached.

The established case presents with persistent, copious, effortless vomiting, the amount of gastric fluid being far in excess of 2 litres per day. The patient complains of thirst, the tongue is dry and the breath foul-smelling. There may be abdominal distension and some tenderness. Auscultation may reveal faint intestinal sounds and small amounts of flatus and even faeces may be passed, but advanced cases have an ominously silent abdomen associated with paralytic ileus.

Radiography of the abdomen will show the enormously dilated stomach. Often dilated loops of small intestine are seen as well and, since in advanced cases the condition merges into distal ileus, the entire gastro-intestinal tract may be involved.

Paralytic ileus due to peritonitis, typhoid fever, diabetic ketosis and uraemia must always be excluded since they require specific treatment apart from fluid and electrolyte replacement. There are also a number of conditions which may cause a so-called 'reflex ileus' but it is unlikely that they will be confused with acute gastric dilatation.[22] Finally it must not be forgotten that mechanical intestinal obstruction may terminate in paralytic ileus.

Symptoms and signs of hypokalaemia, apart from gastro-intestinal paralysis, are rarely encountered, and indeed it is still doubtful whether they can be attributed directly to the lack of potassium.[23, 24] Apathy and listlessness, muscular weakness and pains in the legs, are those most frequently reported and were present in both of the cases described here.

Examination of the urine is always profitable, and for the country practitioner indispensable. The volume, specific gravity and chloride content can all be measured clinically and provide information about the degree of salt and water depletion. When employing Fantus' test for urinary chlorides it must be remembered that in the presence of adrenocortical activity the values may be abnormally low owing to NaCl retention. There is no easy test for urinary potassium, but in the established case of post-operative ileus 1·5—3 g. of potassium may be lost each day.[6]

The electrocardiographic changes attributed to intracellular K^+ deficiency are not as reliable as formerly believed and may be absent in cases with undoubted K^+ depletion.[25] Those most often seen are prolongation of the Q-T interval, flattening or inversion of the T wave, and depression of the ST segment.

Estimations of the plasma K^+ concentration are useful when available but by no means indispensable, nor always reliable, as an indication of K^+ deficiency. It is quite possible to find a normal plasma K^+ concentration in the presence of a marked intra-cellular deficiency. Estimations of cellular K^+ have been made[6, 11] and in all probability the technique will eventually be perfected.

Of far greater practical importance is the frequent, if not invariable, occurrence of hypochloraemic alkalosis in acute gastric dilatation, because it is always accompanied by K^+ deficiency. Biochemical estimations of the degree of alkalosis have been found to be less reliable in many cases than clinical observations on the gastric juice.[1] Starr has drawn attention to the changes that occur; they are of the utmost importance: (1) The quantity is increased and exceeds 2½ litres per day. (2) The colour changes from green to yellow and then to brown, the fluid becoming increasingly turbid and foul-smelling. It is vitally important to appreciate that faeculent vomiting in these cases is not due to regurgitation of intestinal contents into the stomach as a result of a low mechanical obstruction, and certainly not to a gastrocolic fistula. This point is stressed as it has been found that these are precisely the conditions commonly linked to the term 'faeculent vomiting'.

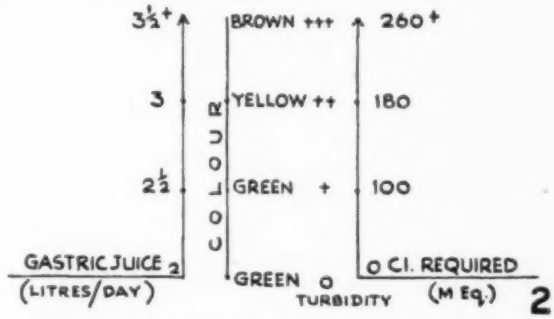

Fig. 2. Correlation between gastric juice and chloride requirements.

The chloride requirements have been correlated to these changes, as shown in Fig. 2.[1] Fantus' test can also be used to estimate the chloride loss in the gastric juice.

This hypochloraemic alkalosis is resistent to NaCl administration and will not be reversed unless the accompanying K^+ depletion is made up too.

TREATMENT

Once the condition has been recognized treatment must be prompt and bold. A delay of a few hours may result in death from oligaemic shock. Timid fluid and electrolyte replacement only delay the fatal outcome, and saline infusion without K^+ replacement increases the K^+ depletion, prolongs the state of ileus and results in circulatory failure or pulmonary oedema. Case 1 will always serve as a bitter reminder of this fundamental rule.

Serum and Dextran may be used to combat shock in the early stages but this must be followed as soon as possible by gastric aspiration and parenteral administration of fluid and electrolyte.

When calculating fluid requirements it must be remembered that an obviously dehydrated patient has a deficiency of at least 4 litres[22] and this must be made up in addition to the requirements for each day. Accurate fluid replacement is impossible without a meticulously kept chart recording the daily intake and output.

Correction of the Cl^- deficiency is guided by the changes in the gastric juice (Fig. 2) and is effected mainly by the use of KCl in order to correct the K^+ deficiency at the same time. If it is felt that this will provide too much K^+, part of it may be given in the form of NH_4Cl.

Describing the treatment of a case of post-operative gastric ileus Logan[9] states that 'rapid estimations of serum potassium are necessary, such as may be obtained with a flame photometer, and it is essential to check the serum levels daily or more often'. Fortunately for the country practitioner clinical observation and bedside tests can provide almost all the information he may need as long as he remembers that K^+ and Cl^- administration go hand in hand in the correction of the alkalosis. Parenteral administration of K^+ must be further guided by the following rules:

1. There must be a good output of urine.

2. K^+ replacement should not exceed 40 mEq. in 3 hours and the replacement solutions should contain not more than 40 mEq. K^+ per litre. (Note that 1 g. of KCl contains 13·4 mEq. K^+.) In the early stages it is best given in the form of KCl added to a solution of 5% dextrose in water, but once the initial deficiency has been corrected and the gastric juice regains its normal character all the bulk ions should be provided by giving a solution such as Darrow's, which contains 4 g. of NaCl, 2·7 g. of KCl, and 52 ml. of molar sodium lactate per litre. As a rule 1 litre of Darrow's solution per day will provide all the electrolyte requirements.

3. ACTH or cortisone should not be given.

4. Oral administration must replace parenteral administration as soon as possible. Here again Darrow's solution may be used.

The change-over to oral feeding is difficult and requires caution even when gastric motility has been re-established. Intravenous KCl administration must not be stopped until recovery is complete, but the amount is gradually decreased as the oral route is developed.

The clinical signs heralding recovery are easily recognized. The quantity and the character of the gastric juice return to normal; signs of dehydration disappear and the tongue stays moist; peristaltic bowel sounds return and flatus is passed regularly; the patient develops an appetite, becomes alert and active, and the days of hopeless despair are replaced by confidence and unbounded optimism.

SUMMARY

Acute dilatation of the stomach, though not often seen in general practice, is an important and serious condition. Two cases, encountered during the last year, are described. They illustrate most of the important features of the condition, as well as some of the problems in fluid and electrolyte therapy peculiar to a country practice.

Etiology, pathogenesis, diagnosis and treatment are discussed in the light of recent knowledge about the role of potassium in gastro-intestinal paralysis.

The character of the gastric juice is a reliable index of the degree of alkalosis accompanying the condition. This hypochloraemic alkalosis is resistent to sodium chloride administration and will not be reversed unless potassium is given too.

Although the emphasis throughout is on potassium deficiency, it must be realized that this is but one part of a generalized metabolic disturbance, every aspect of which should be considered during treatment.

I should like to express my thanks to Dr. A. Cohen for allowing me to attend his patients; to the Matron of the Gordonia Hospital for her willing cooperation during the treatment of these patients; and most of all to the nursing staff, whose careful attention under trying conditions dispelled most of the anxieties associated with prolonged intravenous-fluid therapy.

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ASSOCIATION NEWS : VERENIGINGSNUUS

PHILOSOPHY AND MEDICAL PRACTICE

At the Annual General Meeting of the Transkei Branch, held at Umtata on 5 February 1955, Dr. F. J. Wiles, of Tembuland Hospital, Umtata, delivered the following address.

I

Doctors, more than most people, come into frequent contact with the realities of life. We see people born and we see them die. We see them when they are faced with pain and illness, and we see their varied and sometimes astonishing reaction to the troubles which they have to meet. We have ample opportunity to observe the vagaries of human behaviour.

It is therefore inevitable that we should sometimes ask ourselves what the meaning is of human life and of the universe in which it is contained. No one who thinks a little can avoid wondering what may be the reason behind it. Sometimes we want to cry out in despair, 'What on earth is this complicated business of living all for?'

Before going on to try and answer some of these queries I want first to establish one preliminary point. When one speculates on vague questions such as these one is immediately faced with the difficulty that they are not susceptible to proof of any kind. With the scientific training which we receive nowadays we are apt to be suspicious of anything that cannot be proved. Yet it is remarkable what a lot of things we accept as established facts without any proof whatever. If we look at a picture by a famous artist, we know that it is beautiful and yet it is impossible to prove that it is so. If we hear a Beethoven sonata we accept its beauty as a matter of course, without requiring any proof.

One can also look at more scientific matters and still find proof lacking. Even mathematics, the most exact of all the sciences, is really founded on an assumption. The axiom that things equal to the same are equal to each other is only an assumption for it cannot be proved. Yet without it all mathematical proof falls to the ground. Lest you think this rather far-fetched let me say something further about geometry in particular. The geometry that we all learned at school was that of Euclid, which is a logical system of thought and for many centuries was accepted as being proved beyond all doubt. Yet the modern mathematicians have invented other systems of geometry, because under certain conditions in space Euclid has been found to be invalid. Euclid proved clearly that the 3 angles of a triangle are equal to 2 right angles, yet there is another geometry which proves that they are less than 2 right angles and still a third geometry which proves that they are greater than 2 right angles. All these contradictory systems can be proved, and are equally valid under different conditions.

It seems therefore that scientific proof is not so important as one might suppose. This has been well put in one of Tennyson's poems:

> 'Thou canst prove the Nameless, O my son,
> Thou canst prove the world thou movest in.
> Thou canst not prove that thou art body alone,
> Thou canst not prove that thou art Spirit alone:
> Thou canst not prove that thou art both in one.
>
> Thou canst not prove that I who speak with thee
> Am not thyself in converse with thyself.
> For nothing worthy proving can be proven
> Nor yet disproven. Wherefore thou be wise,
> Cleave ever to the sunnier side of doubt.'

II

The main part of my subject will be divided into two portions. The first is of only theoretical interest while the second has perhaps some practical bearing on therapeutics. The first question that I want to put is this: 'Is the universe just a haphazard affair not moving in any particular direction, or is there some definite purpose and meaning behind it?' My answer is that there must be a driving purpose controlling the destinies of the universe in general and of mankind in particular. The universe came into being for a definite reason, and throughout its long history there has been a guiding hand with a purpose for everything that has occurred. This is the doctrine of Teleology, and I think such a belief in Purpose is the most fundamental faith that man can have. I do not intend to consider the nature of this guiding purpose or even to try and say whether we can identify it with God; that would be venturing into the domain of Theology and Religion which are outside the scope of this address.

Let me however, give two arguments in favour of such a belief. If we look at some intricate man-made apparatus such as a modern X-ray set we are amazed at the ingenuity of the person who designed it. Now it is barely possible that if one took a scrapheap of bits of metal, glass, cord, etc., and threw it violently into the air, it would by chance fall together in the shape of an X-ray set that really worked. Such a result however, is so unlikely that no one would seriously believe that an X-ray set could come into being by chance, without a master-mind to build it. If then we look at the much more wonderful and complicated design of the universe it is surely equally inconceivable that it came into being and keeps going without a master-mind which created it and guides it. When I speak of a Mind having created the universe I do not, of course, mean anything like the human mind; such a Mind must be of a nature quite beyond human comprehension.

My second argument lies in the fact of Evolution. Darwin's original explanation of Evolution, viz. Natural Selection, was strongly supported during the last century, but is not believed in by many biologists today. Most scientists now hold that Natural Selection is quite inadequate to explain the process of Evolution and many go so far as to say that there appears to be some driving force behind the age-long process of development. They cannot of course say wat this force is except that it seems to be something more than ordinary natural laws. It is of interest to note that one of the strong supporters of this theory was the late Dr. Robert Broome, the famous South African palaeontologist. He wrote a book explaining this idea about 30 years ago.

If we accept this belief in purpose we must carry it to its logical conclusion and believe that there is a good reason for everything that exists in the universe. There is reason for life and death and there must also be reason for disease. The fact that we cannot see the reason for parasites, disease-causing bacteria, or cancer cells does not mean that no good reason exists. Look again at the similie of the X-ray apparatus. The layman who examines the intricate mechanism, cannot know the reason for each separate part, but it is obvious to him that each part must serve a useful purpose. The master-mind which was capable of designing such a mechanism would certainly not have included parts that were unnecessary or harmful to the apparatus as a whole. Similarly, it is inconceivable that the guiding purpose behind this marvellous universe should have created anything in it which served no useful

purpose. The reason for parasites and cancer cells may be in the remote past or in the dim future, but it is surely there.

Now if we take this argument a step further, we may find that our position as doctors becomes rather questionable. Is it not possible that our well-meant efforts to treat disease are simply interfering with the purposes of nature? Taking a long-term view of human destiny, do death and disease really matter after all? To us they are major tragedies, but this may be only the limited, short-sighted outlook of the present era. A later age may see things in better perspective, and come to regard such matters quite differently. In the long run will the work of doctors prove to be of much importance?

These questions are rather unpractical, but at least it is fair to say that we as a profession have a grossly exaggerated idea of our own importance. We are too apt to believe that we are great benefactors of mankind, and that human happiness depends more on our profession than on any other. This is, in fact, quite untrue. With the exception of a few doctors whose work prevents the more formidable epidemic diseases, the whole medical profession could be abolished with very little effect on the human race as a whole. A few individuals would die who might otherwise be saved; a few would suffer pain who are now relieved, but the human race would still survive. The treatment of sickness is not one of the basic necessities of life, for man existed and multiplied for thousands of years without therapeutics as we know it today.

There are 4 things which are essential for human life: food, clothing, houses and fuel. If the workers who produce these commodities all stopped work, the human race would soon die out, but it could get along fairly well without doctors. It is only the emotion and sentiment attached to sickness that create the illusion that doctors are indispensable.

This is a melancholy and very humbling thought, but fortunately there is a crumb of comfort for us. If the doctrine of Teleology is universally true, it must be conceded that there is even a purpose for having doctors in the world. We also are cogs in the vast machine, with some small part to play, some slight service to perform.

III

The second part of my subject poses another question: 'What is the real nature of the universe? What is the fundamental stuff of which it is made?' Three main answers are possible. Firstly, the ultimate reality may be only matter—this is materialism. Secondly it may be only mind—this is idealism. Thirdly, both mind and matter may be realities interacting with each other. Philosophers have debated this question since the time of Aristotle, and it is, of course, a problem to which there can never be a satisfactory solution. You will remember the lines from Omar Khayyam:

> 'Myself when young did eagerly frequent
> Doctor and Saint, and heard great argument
> About it and about, but evermore
> Came out by the same door as in I went.'

Yet the fact that there can be no answer will not prevent men from speculating, wondering and arguing; so let us venture to speculate.

I am going to suggest that all material things are an illusion, and the only real basis to the universe is mind or, if you prefer it, thought, which is the primary function of the mind. The old philosophers argued it out in this way: A thing does not exist entirely on its own. It always exists in relation to some other thing. In other words a thing can only have significance in so far as it exists for something else or has meaning for something else. Now it is obviously impossible for one material object to have meaning for another material object. It can only have meaning for a mind which thinks about it. Therefore matter has no significance except when it is in relation to mind or thought; and so it can be said to be unreal. Mind is the only reality, for only it can think about itself or about other objects.

This abstract argument is perhaps not very convincing, but in this era the physicists have given us more definite evidence regarding the illusory nature of matter. Let us look first at space and time in which matter is contained. According to Einstein's Relativity Theory measurements of distance and time vary according to the speed of the observer. Two observers measuring the same events may get entirely different results. To one observer two events may seem to occur simultaneously and at the same place. To another observer the same two events may appear to be widely separated in time and space. In other words, time and space are not hard-and-fast measurements, but purely relative concepts.

Next we must consider matter itself. Matter used to be regarded as very concrete stuff, and one of its fixed properties was that the mass of any given quantity of matter remains constant. (For the sake of simplicity I will speak of 'weight' instead of 'mass' although the two terms are not strictly synonymous.) It has been found that the weight of an object is not constant, but increases with increasing velocity. At ordinary speeds the difference is negligible, but at one-half the speed of light the weight of an object is increased by 1/7th, and at 9/10ths of the speed of light it is 2½ times as heavy. This extra weight is due to the energy which the object derives from its velocity. It therefore appears that matter simply consists of energy. The same conclusion can be arrived at by considering the structure of the atom. The protons and electrons which make up an atom used to be thought of as minute particles of matter until it was found that they do not behave like particles. The only way in which their behaviour can be explained is to regard them as waves of energy. Thus matter in the conventional sense of the word does not really exist.

These considerations make it not unreasonable to believe that the material universe is an illusion, and that the only reality is mind or thought. This concept has a deep significance for those who practise medicine. We need to grasp firmly the idea that in dealing with our patients we are dealing chiefly with minds. The study of disease must not restrict our vision so that we fail to see further than the physical body. The mind is the real person.

MEDICAL PRACTICE

With our narrow scientific outlook we are apt to think of a patient as an ulcer or an anaemia or an ingrowing toenail. We must see each patient as a person with thoughts and memories, longings and anxieties, happy or unhappy family relationships, and all the other complexities which make up the personality. He is a person who is worth getting to know, and what he thinks about is more important than what happens in his body.

There is no doubt that doctors today have not the respect and confidence of the public to the same extent as in previous generations. Patients are often suspicious and critical. Our grandfathers were much less successful in treating disease, yet their patients believed in them almost with reverence. They lacked our scientific equipment, but they had the gift of getting to know patients and understanding their problems. Something of that art has been lost, with the result that patients often feel they are not getting a square deal. I am not referring to bad diagnosis and treatment, but to matters which even a conscientious and efficient doctor may overlook.

The complex doctor-patient relationship cannot be analysed in a few sentences, but I will suggest briefly three ways in which patients could be handled more sympathetically. Firstly, the patient often does not get a clear and rational explanation of his condition. Most patients have intelligent minds and they like to be told exactly what is going on in language that a layman can understand. They should not be put off with vague pseudo-scientific terms whenever we do not know what is the matter with them.

Secondly, the financial struggle which most patients have is not fully realized. It is unfair to let patients with a small income pay for treatment if a free service is available, or to order costly diagnostic and therapeutic procedures which are not essential. Too often an exorbitantly expensive 'mycin' or vitamin mixture is prescribed when a cheaper treatment would suffice.

Finally, there is a tendency, if no organic disease is found, to dismiss the patient's complaints as being due to hysteria, hypochondria or some Freudian inhibition. This diagnosis satisfies the doctor, but has never yet cured a patient. Such a patient goes from doctor to doctor and gets a variety of treatments but no relief. He comes to be regarded as a subject for psychotherapy, when all he required to start with was a doctor with an insight into the human mind and its needs—a doctor who can understand.

This can all be summed up in a sentence from a recent article in the Lancet: 'Where the science of the human body ends, that of the soul begins.'

IN MEMORIAM

GEORGE KEBLE MOBERLY, L.R.C.P. & S. (EDIN.), L.F.P.S. (GLAS.)

The death took place in Durban in December last year of Dr. G. K. Moberly, of Kloof, Natal.

George Keble Moberly was born in 1872 at Bonn, Germany, where his father was the British chaplain at the University. Educated in Scotland, he took the Triple Qualification in 1894 and immediately emigrated to Natal. He practised in Ladysmith and Dundee districts before the South African War, and during that war and the East Africa Campaign of the First World War he served as an army medical officer.

From 1902 until 1912 he was district surgeon of Eshowe, and thereafter until his retirement in 1934, district surgeon of Empangeni in Zululand. Of his experiences among the Zulus he published two books of short stories, *Zululand Romance* and *Square Deal*.

Dr. Moberly was licensed by the Natal Medical Committee in 1894, and—as far as can be ascertained—was the senior practitioner on the register in South Africa.

HERBERT JOHN ORFORD, O.B.E. (MIL.), M.B., B.CH. (BIRM.), M.R.C.S., L.R.C.P.

Dr. H. J. Orford died on his farm at Klerksdorp, Transvaal, at the age of 77. He had retired from civilian medical practice over 30 years ago, but served as A.D.M.S. in the second world war, as he had done in the first. Dr. Orford was founder (more than 30 years ago) and Past-President of the Sussex Cattle Breeders' association, and member of the Executive Committee of the Witwatersrand Agricultural Society for many years.

Major-General A. J. Orenstein, C.B., C.M.G., C.B.E., *M.D., LL.D., F.R.C.P., writes as follows:* I first met Orford in 1917. He was then A.D.M.S. Lieut.-Colonel in charge at Kimberley. He was an outstanding officer. He was awarded the Military O.B.E. Unfortunately he contracted pneumonia there and nearly died. He practised in Klerksdorp with the late Dr. H. E. Brawn. They were held in very high esteem as practitioners and as men of outstanding integrity. Orford became interested in cattle breeding, and retired from practice in the early twenties.

In the second war he again joined the S.A.M.C. He served for a while in the Middle East campaign, but was most of the time in Sonderwater Camp, where he was A.D.M.S. with the rank of Colonel. He was promoted to Brigadier on relinquishing his appointment.

Orford was the very prototype of honesty, integrity and forthrightness. He despised anyone who did not live up to his standard, and he never pulled his punches in commenting on any person, no matter what his status, whom he deemed dishonest or unfair. There are few like him.

CONFERENCE ON BIOFLAVONOIDS AND THE CAPILLARY

Dr. Albert Szent-Georgyi, director of the Institute for Muscle Research at Woods Hole, Mass., at a conference on Bioflavonoids and the Capillary, held on 11 February 1955 and sponsored by the New York Academy of Sciences, announced the isolation of a new flavonoid-like substance from calf thymus. It was also announced that flavonoids, which counteract capillary fragility, have been used with good results in the treatment of acute poliomyelitis and for cases of habitual abortion.

The newly discovered thymus substance is present in the high concentration of 0·1 mg. per g. of tissue. Hitherto it has only been found in plants, and has resisted discovery in animals since it exists in a colourless form as part of a complex in the thymus.

Dr. Szent-Georgyi suggested that it might provide clues to the secret growth, normal and abnormal. Since the thymus grows until the age of 2 years and does not atrophy until after puberty, he speculated whether the new flavonoid might be a hormone necessary to normal growth, and whether abnormal growth might result from its absence, because smaller amounts of it might be required even after general growth has stopped. Here, he thought, is a possible explanation why cancer and other chronic diseases are most common after the thymus atrophies.

In another report at the same conference, Dr. Robert Greenwalt stated that more than 80% of a group of women with histories of abortion were found to suffer from abnormal capillary fragility. Seven patients with records of 3-8 abortions were given hesperidin (a flavonoid) and ascorbic acid; 4 were delivered of live infants. Of 13 patients, with 2 previous abortions each, 11 were delivered of live infants. The available statistics indicated that the expected rate would have been only 1 baby in the 1st group and 8 in the 2nd.

Dr. George J. Boines also reported that hesperidin, combined with ascorbic acid, had proved effective in treating a group of 400 patients with acute poliomyelitis, all of whom showed abnormal capillary fragility. Of these 80% responded in an average time of 5 weeks. Appetites improved within the 1st week, and by the 2nd week there was an increased warmth to touch in the involved extremity. He concluded that 'patients responded better and more quickly when there was an improvement in capillary function'.

PASSING EVENTS : IN DIE VERBYGAAN

Union Department of Health Bulletin. Report for the 5 days ended 29 March 1955.

Plague, Smallpox: Nil.

Typhus Fever, Cape Province: One (1) Native case in the Queenstown district. One (1) Native case in the Willowvale district. Diagnosis of both cases based on clinical grounds only.

Epidemic Diseases in Other Countries:

Plague: Nil.

Cholera in Calcutta, Pondicherry (India); Chalna, Chittagong, Dacca (Pakistan).

Smallpox in Kyaukpyu, Moulmein, Rangoon (Burma); Phnom-Penh, Ahmedabad, Allahabad, Alleppey, Bombay, Calcutta, Cochin, Delhi, Jodhpur, Kandla, Kanpur, Lucknow, Madras (India); Dacca, Karachi, Lahore (Pakistan); Nhatrang (Viêt-Nam); Mogadiscio (Somalia); Tanga (Tanganyika).

Typhus Fever: Nil.

Dr. Arthur E. Amoils, M.B., B.Ch. (Rand), D.L.O. (London), F.R.C.S. (Edin.), has joined Drs. H. Penn and J. Fine in Ear, Nose and Throat practice at 709 Medical Centre, Jeppe Street, Johannesburg. Telephones: Rooms, 23-2334; Residence, 43-3546.

* * *

Faculty of Medicine, University of Cape Town. The following paper is due to be read at Research Forum on 4 May 1955 at 12 noon in the large A Floor Lecture Theatre, Groote Schuur Hospital. The speaker will be Mr. D. J. du Plessis. The subject will be *Swellings of the Parotid Glands (Excluding Mixed Salivary Tumours)*.

* * *

Memorandum on the Outbreak amongst the Nurses at Addington Hospital, Durban. Mr. R. C. J. Hill, F.R.C.S.E., over Chairman

of the Clinical Section of the Investigating Committee, Durban, under whose name this article was published in the issue of 9 April 1955 (page 344), supplied the names of the members of the Clinical Section, and suggests that they should be published. They are: Dr. T. M. Adnams, Dr. M. Casson, Dr. R. W. S. Cheetham, Mr. R. C. J. Hill, Mr. C. J. Kaplan, Dr. B. V. Schulze and Dr. H. L. Wallace.

* * *

General practitioners are advised that the Workmen's Compensation Commissioner has agreed to pay fees for X-ray examinations carried out by general practitioners, provided that the accounts are endorsed: 'It was in the best interests of the workman that he should have been X-rayed at the time'.

* * *

Erratum: In Dr. H. T. Phillips' letter in the *Journal* of 9 April 1955 at p. 351, the word 'successively' (in the 23rd line of p. 352) was written in error as 'successfully'. The sentence should read 'A middle-aged widow with an unhappy childhood and 5 small children had her gall-bladder, appendix, uterus, coccyx and patella successively treated by operation'.

* * *

The 7th International Congress of Comparative Pathology will be held at Lausanne from May 26 to 31. Scientific meetings to be followed by discussions will be held when papers will be presented on Viral Diseases transmitted to Man by Animals, Atmosphere Pollution and Growth Disturbances in Comparative Pathology. A full programme of social activities has been planned to follow the Congress.

* * *

New International Maritime Convention. A maritime Convention of the International Labour Organization requiring all persons employed on sea-going vessels to be medically examined before being signed on and periodically thereafter will come into force in August 1955. It will, however, not apply in the United Kingdom or the Union of South Africa, which are not ratifying countries. The ratifying countries include Canada, France, Italy, Norway, Portugal, etc.

The Convention applies 'to every sea-going vessel whether publicly or privately owned which is engaged in the transport of cargo or passengers for the purpose of trade'. It does not apply to vessels of less than 200 tons gross register, fishing vessels, estuarial craft, or certain wooden vessels of primitive build.

The earlier maritime Convention (1921) required medical examination for young seafarers only.

Of the international labour Conventions adopted by ILO since 1919, 25 or nearly one quarter referred to seafarers.

Erratum—An error occurred in the paragraph headed 'Unregistered Practitioners' in the Report of the Meeting of Federal Council which was published in the *Journal* of 2 April 1955 (page 324). The proposal that in future no benefit or medical aid society which permits payment to unregistered practitioners or auxiliaries was not passed. The first sentence of the paragraph is therefore wrong and should be deleted. It was decided to seek a ruling on the subject from the Medical Council and the second paragraph is therefore correct.

* * *

Fourth Commonwealth Health and Tuberculosis Conference, London, 1955. The Duchess of Kent, President of the National Association for the Prevention of Tuberculosis, will attend the Association's Fourth Commonwealth Health and Tuberculosis Conference at the Royal Festival Hall, London, on 22 June, and welcome the delegates.

The Conference will be opened on 21 June, by the Rt. Hon. Iain Macleod, M.P., Minister of Health, when the subject will be 'The Preventive Outlook Today'. The Rt. Hon. Alan Lennox-Boyd, Secretary of State for the Colonies, will speak at the session on 'Tuberculosis—A Problem of Different Races', and the Rt. Hon. Viscount Swinton, Secretary of State for Commonwealth Relations, will also attend the Conference and address the delegates. There will be discussion on 'Child Hygiene and Infection', 'Tuberculosis in Industry', 'Differential Diagnosis', and 'Tuberculosis and Leprosy'.

Conference speakers will include Dr. Johannes Holm (World Health Organization), Dr. P. V. Benjamin (India), Prof. Etienne Bernard (France), Dr. Walsh McDermott (U.S.A.), Dr. E. Ross (Canada), Dr. H. G. Trimble (U.S.A.), Prof. Arvid Wallgren (Sweden), and Sir Geoffrey Marshall, Dr. George Day, Dr. Honor Smith and Mr. J. L. Martin, Architect to the London County Council.

The Exhibition will be opened by the Rt. Hon. James Stuart, Secretary of State for Scotland. Cinema films will be shown. Visits to well-known hospitals and sanatoria will be arranged for delegates. The Lord Mayor and Corporation of London are giving a reception for delegates, and other receptions are being arranged by medical associations and societies.

Representatives will attend the Conference from: Aden, Australia, Belgium, Bermuda, British Guiana, British Solomon Islands, Canada, Ceylon, Cyprus, Finland, France, Gambia, Germany, Greneda, Holland, Hong Kong, India, Indonesia, Kenya, Leeward Islands, Malaya, Mauritius, Nigeria, Northern Rhodesia, Norway, Sarawak, Sierra Leone, Singapore, South Africa, Southern Rhodesia, Spain, Sweden, Switzerland, Tanganyika, Trinidad, U.S.A., Yugoslavia.

NEW PREPARATIONS AND APPLIANCES : NUWE PREPARATE EN TOESTELLE

MERCLORAN AND MERCARDAC—*Two new mercurial diuretics.* P.D. and Co. (Pty.) Ltd., the South African subsidiary of Parke Davis and Company, announce the introduction of their oral mercurial diuretic 'Mercloran' together with 'Mercardac'—a parenteral form, and supply the following information. This diuretic has acquired a high reputation on account of its lack of toxicity, ease of administration and stability.

Mercloran: Mercloran (chlormerodrin, Parke Davis, 3-chloromercuri-2-methoxy-propylurea) is an effective *oral* diuretic which is administered daily. Its use obviates 'the see-saw effect of oedema and dehydration' which may often occur on a regimen of intermittent treatment.

Recent comparative assays of 8 *oral* diuretics including mercurial, xanthine and amino-uracil diuretics, showed Mercloran to have the highest rating. This rating was derived from the ratio between 'diuretic effectiveness' and 'corresponding incidence of gastro-intestinal irritation'. This rating ranged from ammonium chloride (52—the lowest) to Mercloran (284—the highest).

The diuretic effect of Mercloran approaches the efficacy of the most efficient parenteral mercurials whereas other oral diuretics come no nearer than a fourth of the diuretic potency of the standard. It is well tolerated. Nausea and diarrhoea have occasionally been observed with a high dosage schedule but these disappear on withdrawal of the drug after which therapy is recommended on a lower dosage schedule.

Owing to the daily sustained diuretic action of Mercloran, its use often permits modification or withdrawal of dietary salt restriction thus avoiding the hypochloraemia resulting from the massive diuresis produced by the intermittent parenteral mercurial diuretics.

Indications. Mercloran is indicated in the treatment of congestive heart failure, cardiac asthma, cardiac dyspnoea, recurring oedoema and ascites, fluid retention masked by obesity and patients averse to a low salt diet.

Contra-Indications. As with all mercurials Mercloran is contra-indicated in acute nephritis and other forms of severe renal damage. The average daily dose ranges from 1-4 tablets (equivalent 10-40 mg. of mercury), which may be increased to 6 tablets, according to the severity of the oedema.

Package Information. Tablets, each containing 18·3 mg. chlormerodrin N.N.R. equivalent to 10 mg. of mercury in bottles of 25 and 250.

MERCARDAC. Mercardac (meralluride sodium U.S.P.) is a parenteral mercurial diuretic. Each c.c. represents the sodium salt of meralluride (methoxyoxy-mercuripropyl-succinyl urea with theo-

phylline) equivalent to 39 mg. of organically combined mercury with 48 mg. of theophyllin.

Indications and Contra-Indications. As for Mercloran.

Dosage. From 1 to 2 c.c., equivalent to 39-78 mg. of mercury, depending on the condition of the patient and the route and frequency of administration. An initial dose of 0·5 c.c. or less is recommended in order to ascertain the responsiveness of the patient and to detect occasional idiosyncrasy. The dose for children is in proportion to body-weight. The interval between injections may vary from 1 day to 1 week, according to the individual patient's requirements.

Routes of Administration. The intramuscular route has almost completely taken the place of the intravenous injection. Mercardic is less irritant to the muscle and less painful than most other preparations.

Side-reactions and Toxicity. At average dose levels, Mercardac is well tolerated. With doses of more than 2 c.c. daily, however, symptoms of gastro-intestinal upset may be encountered. Patients have received Mercardac 2 or 3 times a week for up to three years without evidence of renal damage.

Package Information. Available in boxes of 6×1 c.c. ampoules, boxes of 12×2 c.c. ampoules, and rubber-capped vials of 10 c.c.

NUFER. British Drug Houses (S.A.) (Pty.) Ltd. supply the following information:

Each tablet contains ferrous sulphate, exsiccated, 200 mg. (3 gr. approx.), ascorbic acid 10 mg. and acetomenaphthone 2 mg.

Nufer is indicated in the treatment of hypochromic anaemias. The preparation is well absorbed and, in addition to providing a most effective form of iron therapy, does not give rise to gastro-intestinal disturbances.

Dosage. Children 1 tablet 2 or 3 times a day. Adults 2 tablets 3 times a day. Nufer is best taken during or immediately after meals.

It is available as sugar-coated tablets in bottles of 50 and 500.

Reprints of Gillhespy's paper entitled 'The treatment of hypochromic anaemia with a compound containing ferrous sulphate, vitamin C and K' (The Medical Press, 3 February, p. 112) and trade literature are available on request.

NEO-GELICIL EYEDROPS. Saphar Laboratories Limited supply the following information about their new product Neo-Gelicil Eyedrops:

The eyedrops contain the antibiotic Neomycin (5 mg. per c.c.) and sodium propionate (5%). These antimicrobial agents are effective against practically all bacterial and fungal infections of the eyes, including the Koch-Weeks bacillus, causative agent of 'pink-eye', Bacillus pyocyaneus and Pityrosporum ovale, a yeast-like organism which is found in chronic seborrhoeic blepharo-conjunctivitis.

Most, if not all, penicillin-resistant strains of staphylocci will respond to the combined action of Neomycin and sodium propionate.

Neo-Gelicil Eyedrops are stable for at least one year and require no refrigeration. Available in ¼-oz. dropper bottles.

BOOK REVIEWS : BOEKRESENSIES

CLINICAL EXAMINATION OF THE NERVOUS SYSTEM

Clinical Examination of the Nervous System. By G. H. Monrad-Krohn, M.D., F.R.C.P. Third Edition. Pp. 428+xx. With 165 illustrations. 36s. London: H. K. Lewis & Co. Ltd. 1954.

Contents: 1. Anamnesis. 2. Status Praesens. 3. Mental State. 4. Cranial Nerves. 5. Articulation. 6. Head. 7. Spine. 8. The Motor System. 9. Sensory System. 10. Reflexes. 11. Trophic And Vasomotor State. 12. The Standing Position. 13. The Gait. 14. Electrical Examination. 15. Examination of Cerebro-Spinal Fluid. Appendix. I—XII. Index.

How pleasant it is to welcome an old friend—slightly more corpulent, with a little more experience added to the great wisdom that was always present, and still ever youthful in outlook and enthusiasm! In the course of a professional life there are some books which come early to one's notice and which one handles repeatedly, recommending them first to fellow students and later to one's pupils. As they reappear in subsequent editions one watches, almost with a tinge of personal pride the manifest evidences of their popularity and the direct compliment to the author. Such is this book of Monrad-Krohn's which has gained world-wide fame. Ten editions in 33 years, surely that is all the recommendation any book needs! One can only add that with each edition the author has maintained his original high standard; he has kept pace with every advance in neurology and with all the ancillary methods of investigation; and those who study the book will find that the subject ceases to be the difficult mystery so many still find it. It is a must for every student and for every practitioner who has only an older edition on his shelves.

S.B.

CLINICAL CHEMISTRY FOR TECHNICIANS

Techniques in Clinical Chemistry. By Frederick N. Bullock F.I.M.L.T., F.R.M.S., F.C.S. Pp. 171+ix, with illustrations. 16s. 6d. Bristol: John Wright & Sons Ltd. 1954.

Contents: 1. Fundamentals. 2. Preliminary Preparation. 3. Examination of Blood. 4. Cerebrospinal Fluid Analysis. 5. Analysis of the Faeces. 6. Gastric Analysis. 7. Examination of Urine. 8. Tests of Renal and Hepatic Function. Appendix. Index.

This book is intended to guide recruit technicians in clinical biochemistry. The first part of the book deals with 'fundamentals', viz. volumetric analysis, standard solutions, buffers, indicators, valency, colorimetry, pH. etc. This part is sketchy, unsatisfactory and opinionated. In laboratory manuals it is unusual, to put it mildly, that photo-electric colorimeters and spectrophotometers should be relegated to 'research for estimating very minute concentrations of certain substances'.

The technique of the simple tests described is fairly well set out, but the phrase 'unpublished experiments of the author' appears far too often to be convincing. The text abounds with inaccurate and confused statements e.g. 'the finding of glucose in the urine should automatically lead to the estimation of the blood sugar, preferably by means of the glucose tolerance test'.

In the section on cerebrospinal fluid one finds the statement that a total protein of 30 mg. % is 'slightly, but definitely pathological'. What does that mean? In the same section one finds that 'certain *virus* infections such as *typhus* almost always cause a protein increase'.

Potassium is dismissed in grand manner with the following: '....estimations (of potassium) are more of academic interest than of practical applicability'.

Only one reference is given, and that one is given differently in the text and in the footnote.

There would appear to be no real need for this book unless it were drastically re-edited.

A.A.K.

PELVIC OPERATIONS

An Atlas of Pelvic Operations. By Langdon Parsons, M.D., and Howard Ulfelder, M.D. Pp. 231 with 197 plates. £7 13s. Philadelphia: W. B. Saunders Company. 1953.

Contents: Section I. Abdominal Operations. 1. Considerations Preliminary to All Abdominal Surgery. 2. Helpful Hints for Hysterectomy. 3. Modifications of Total Hysterectomy. 4. Conservative Operations. 5. Operations on Tube and Ovary. 6. Operations on Tube and Ovary. 7. Operations Involving the Intestine. 8. Operations Involving the Urinary Tract. 9. Operations on the Abdominal Wall. 10. Complications Following Abdominal Operations. *Section II. Vaginal and Perineal Operations.* 11. Considerations Preliminary to all Vaginal and Perineal Operations. 12. Vaginal Plastic Procedures. 13. Combined Abdominal and Vaginal Procedures. 14. Operations for Repair of Fistulae. 15. Minor Operations. *Section III. Operations for Malignant Disease.* 16. Operations for Malignant Disease. Index.

This book, as its title indicates, sets out to be a guide to the surgeon who undertakes pelvic surgery. It is not a surgical text-book and the physiological aspects are not considered. It is, however, an excellent book describing the steps of the common pelvic operations. The techniques described are the standard ones used in Massachussets Hospitals under the tutelage of Joe V. Meigs. Excellent diagrams illustrate each step of the operation, doing away with the necessity for lengthy descriptions. The text is lucid and concise, and designed to supplement the illustrations.

The book is divided into 3 sections. The 1st section deals with abdominal operations, and procedures involving the uterus, tubes and ovaries are fully described. In addition a part is devoted to general surgical procedures that the surgeon may have to carry out during the course of pelvic operations. Particular attention is given to the repair of injuries of the bowel and urinary tract. A section also deals with the repair of herniae of the anterior abdominal wall.

The 2nd section is devoted to vaginal and perineal procedures, including operations requiring a combined abdominal and perineal approach. Included in this section are operations for the repair of fistulae and alleviation of stress incontinence.

The 3rd section is devoted to operations for pelvic malignant disease. These operations are designed as curative and are of necessity very extensive. This section reflects the teaching of Joe V. Meigs, particular emphasis being placed on the importance of block dissections where the glands and the affected organ are removed as one piece. This method of attack is technically no more difficult than that of removing the diseased organ first and dissecting the glands out later.

This book is a publication of very high standard both in its content and in its presentation. It would be a valuable addition to the bookshelf of the gynaecologist or the general surgeon whose surgical bent is towards the pelvis.

D.M.

CORRESPONDENCE : BRIEWERUBRIEK

THREE GENERATIONS

To the Editor: During a recent conversation among some of our colleagues, I was asked by Mr. Robert Lane Forsyth if the Fehrsen family could parallel the Forsyth achievement in having three generations of doctors living contemporaneously. The question was no doubt prompted by the fact that during the past four generations our family has had a fair leavening of doctors. I had however to admit that we couldn't equal the Forsyth achievement, which I think must constitute a record in the medical annals of South Africa. It would be interesting to put the question to the readers of the *Journal*.

In the light of the much-discussed and vexed question of patient-doctor relationship, I was prompted to relate the following incident: Shortly after starting practice in Beaufort West 36 years ago I was approached by a fine-looking old Afrikaans retired farmer and asked to attend him. He gave as his sole reason that my grandfather had treated him during his youth and that my uncle had attended him in his middle age. To quote his words, 'Dit is my begeerte dat die derde geslag van jou familie moet nou oor my dokter'. I must admit that I was touched and pleased by this old patriarch's kindly sentiments and gesture.

F. O. Fehrsen

Elsfield
Milner Road
Rondebosch, C.P.
14 April 1955

UNEXPECTED REACTIONS

To the Editor: In view of Dr. Albertyn's letter [1] on unexpected, and serious, reactions, the following case is of interest.

Mr. F., aged 34, an Italian male in good health, of a robust physique, was seen in my rooms complaining of an early web-space infection of his hand sustained in his duties as a butcher. It was decided that he should receive, together with local treatment, several injections of penicillin.

He was given 2 c.c. of an aqueous solution of procaine penicillin, 300,000 units per c.c., in the left deltoid. Within 2-3 minutes of the injection he said that he felt 'peculiar' and had a prickling sensation of the skin in his inguinal regions. His pulse was full, and regular at 70-80 per minute.

In a very short space of time he had become markedly collapsed, with extreme sweating, peripheral vasoconstriction, dyspnoea, and an inability to see, while his conjunctivae were very suffused. He became semi-conscious, and was incontinent of urine, with profuse vomiting a preliminary feature. He received adrenaline and coramine, but his return to normal was slow, and his pulse became palpable after only ¾ hour.

On detailed questioning it transpired that he had, about 1 year previously, received a series of penicillin injections with no untoward effect, although 'M. & B.' on another occasion had resulted in a mild transient rash of the arms and trunk. There is a history of asthma in his father. I feel this may be an example of procaine sensitivity.

J. A. G. Matthews

P.O. Box 112
Scottburgh
Natal
3 April 1955

REFRESHER COURSE FOR G.P.S

Aan die Redakteur: Ek sien die Universiteit Kaapstad hou gedurende die winter hul tweede kursus hierdie jaar, vir algemene praktisyns. Ongelukkig word die getalle baie beperk en kan net 'n sekere getal dit bywoon. Hoe lyk dit met Johannesburg en Pretoria? Kan hulle nie jaarliks of elke 6 maande ook sulke kursusse reël nie? Dit sal dan baie meer algemene praktisyns 'n kans gee om hul kennis te verryk, veral die verafgeleë plattelandse praktisyns.

T. B. de Bruyn

Cradockstraat 4
Steynsburg
4 April 1955

OBSCURE CASES OF DIPHTHERIA

To the Editor These two cases and their conclusions, briefly presented below, may prove of interest to my colleagues.

Atypical diphtheria seen during the course of an epidemic may not catch one napping, but as the initial case (as occurred in each instance below) showed correct diagnosis may well be overlooked until it is too late.

Case 1 (1953). A child of 12 years presented as a 'text-book' example of Ludwig's angina. The swelling under the jaw (midline) was most pronounced, the tongue elevated. and the whole neck brawny, with dyspnoea marked. It was decided to give antibiotics a trial before attempting the comparatively difficult incision and dissection required to drain a Ludwig's infection correctly. On the 2nd day of treatment, membrane became apparent in the throat, and despite heroic appropriate measures the child died in heart failure on the 6th day. Early administration of antitoxin might have been life saving.

Case 2 (1955). A child of 18 months presented with laryngeal stridor and sternal retraction of sudden onset. Careful scrutiny revealed no throat lesion nor any sign of infection whatsoever. The temperature was normal. After a stormy night (for all) hospitalization and X-ray for possible foreign body in the air passages was advised. At this stage the infant became intensely cyanosed and lapsed into semi-coma. A second examination revealed a typical white membrane barely visible, rising from the depths of the posterior pharynx. Emergency tracheotomy performed there and then, and subsequent therapy, permitted the little patient to recover—but only just.

As a general rule for the harassed isolated practitioner, one feels that it is worth remembering:

(i) Any recent generalized swelling of the soft tissues between the upper neck and jaw,

(ii) Any sudden obstruction in the air passages (particularly in the case of young children).

is due to the Klebs-Loeffler bacillus until proved otherwise. Membrane must be sought for repeatedly.

Arnold Rieck

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